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Original Article

CIRCUMSCRIBED HÆMORRHAGIC CORTICAL EN- CEPHALITIS, WITH THE REPORT OF A CASE IN WHICH THE LESION WAS LIMITED TO THE MOTOR ZONE, THE CHIEF CLINICAL MANIFESTA- TION BEING JACKSONIAN EPILEPSY.¹

By CHAS. K. MILLS, M.D.,

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CIRCUMSCRIBED cortical encephalitis studied clinically, and its existence demonstrated by necropsy and microscopical examination, while not unique, is still so rare as to give value to the report of a new case. I shall not go into the history and literature of the subject, which is made unnecessary by the valuable articles of Oppenheim,² Hoppe,³ and others. The entire

¹ Read before the meeting of the American Neurological Association, held in Boston, June 4 and 5, 1906.

² Oppenheim, H., on "Die Encephalitis und der Hirnabscess," vol. ix. of Nothnagel, "Specielle Pathologie und Therapie," Wien, 1897. Oppenheim has a good practical article in his text-book on "Diseases of the Nervous System" on the different forms of acute non-purulent encephalitis.

³ Hoppe, H. H., "Acute Hæmorrhagic Cortical Encephalitis," *Journ. of Nerv. and Ment. Disease*, vol. xxix., No. 9, September 1902.

subject of non-purulent encephalitis, cortical and basilar, has received most attention from German writers, although a few excellent articles have been written by French, English, and American neurologists, that of Hoppe being the most elaborate contribution, so far as I know, of anyone writing in this country.

The impulse given to the study of cortical encephalitis by Strümpell¹ is a good illustration of the value of the application of deductive reasoning in the development of exact knowledge. Strümpell, largely on theoretical grounds and on the basis of analogies, advocated the view that certain of the infantile palsies of children were due to a cerebral polio-encephalitis comparable to the polio-myelitis which had long been the well-known most important cause of the spinal infantile paralysis of children. The truth of his deduction has since been absolutely demonstrated, although as yet only in a limited number of cases.

For the part played by Leichtenstern, Koenigsdorf, Schmidt, Raymond, and others in the history of the subject, the reader is referred to the monograph and to the articles of Oppenheim and Hoppe above cited.

The first case of circumscribed hæmorrhagic encephalitis, cortical and subcortical, with necropsy and microscopical examination, recorded in this country was by Dana and Schlapp² before the meeting of the New York Neurological Society held January 2, 1900. The patient was a man sixty-seven years old, in whose blood the malarial plasmodium was found. After headache he had a hemiplegic seizure. His temperature was at first 104° F., but had subsequently fallen, rising again before death. His chief focal symptoms were left hemiplegia almost complete, immobility of tongue, and marked oral paresis. After he had been in the hospital for a week the paralysis of the arm and leg disappeared, but that of the tongue and lips persisted. No sensory symptoms were noted. An area of hæmorrhagic softening was found in the lower portion of the post-central gyre and in the marginal gyres, involving mainly the deep part of the upper lip of the Sylvian fissure. Two or three smaller areas of hæmorrhage were found in the centrum ovale. These were the

¹ Strümpell, A., *Magdeburger Naturforscher Versammlung*, Sept. 22, 1884.

² Dana, C. L., and Schlapp, M. G., *Journ. of Nerv. and Ment. Disease*, vol. xxvii., No. 2, February 1900.

only brain lesions, but the spinal cord showed two polio-myelitic foci, one in the upper and the other in the lower portion of the thoracic cord. Microscopical examination of the area of cerebral disease showed dilatation of blood-vessels, extravasations of blood, and great proliferation of cells. Small areas of softening were also present in the neighbourhood of the hæmorrhagic extravasations. The appearances closely resembled those described as encephalitis hæmorrhagica. The case had some interesting features in focal symptomatology. The paralysis of the tongue and lips and the site of the lesion pointed to the existence of centres for these parts in the grey matter of the roof of the Sylvian fossa. Dana and Schlapp suggest that the case might be classed under the head of degenerative hæmorrhagic encephalitis. Their examination of the autopsy records of sixty-seven cases of apoplexy showed that forty-two of them were hæmorrhagic, eleven embolic, eleven thrombic, and three encephalitic. The case recorded by them is of interest in connection with that which will be presented in this paper, because of its focal motor symptomatology and the circumscription of the lesion. Jacksonian epilepsy was not present, but cortical or cortico-subcortical paresis and paralysis were marked symptoms.

Harlow Brooks¹ reviews the subject of acute non-suppurative encephalitis in an interesting manner. He believes that a considerable number of cases which recover are regarded as hysteria. The lesions of the disease are enumerated by him as : (1) Cerebro-spinal meningitis, probably secondary to cerebritis and of a "cellular type." (2) General non-septic cerebritis affecting all parts of the cerebrum, and most marked in the cortex and particularly so in that of the motor areas. (3) Degeneration of many of the ganglion cells of the cortex. (4) Degeneration of many of the fibres arising from the large pyramidal cells of the cortex. (5) Diffuse degeneration affecting many of the fibres passing through both internal capsules. (6) Similar changes in the cerebellum. (7) Degeneration of many of the tracts of the pons and oblongata. (8) Degeneration of the chief descending tracts of the spinal cord. (9) Slight changes of the cytoplasm of the ganglion cells of the anterior horns of the spinal cord, probably secondary. The symptoms which caused

¹ Brooks, Harlow, *Medical News*, Aug. 8, 1903, vol. lxxxiii., No. 6.

these conditions in the case reported by Brooks were coma, stertorous respiration, absent patellar reflexes. The unconsciousness persisted for three weeks. Necropsy showed no gross organic lesions.

Cases of acute circumscribed hæmorrhagic cortical encephalitis of the motor zone with Jacksonian epilepsy as a part of their symptomatology are very rare, and yet one of the first cases of this affection recorded had among its symptoms both cortical epilepsy and hemianopsia. This case was recorded by Fürbringer.¹

Batten,² who has discussed the subject of cortical encephalitis in a most interesting manner, especially dwelling upon its relations to polio-myelitis and to encephalitis involving the basal nuclei, has recorded a case with necropsy in which a boy nine years old had aphasia of motor type and right hemiplegia, the eyes and head being turned to the left. Consciousness was at first not lost, but the child soon became drowsy, cried out occasionally, and put his left arm to his head as if in pain. Jacksonian spasm was not present. The coma deepened and the patient died three days after the onset of the disease. Microscopical examination of the motor region showed numerous small hæmorrhages and thromboses of the smaller vessels; but although this condition was most marked in the motor region of the cortex on the left side, it was not limited to that region. In this case, as in that of Fürbringer, the affection was attributed to influenza.

In another case without necropsy, recorded by Batten, a boy two and a half years old, the patient had for a time slow rhythmical movements of the left arm and leg. These later passed off and similar movements occurred in the right arm and leg, the latter persisting but becoming gradually less and less marked. Other focal symptoms were paralysis of the left external rectus, ataxia preventing him from walking, and intention tremor in the right arm and leg. The right knee-jerk was more active than the left; the right plantar reflex gave flexion and the left an extensor or Babinski response. This patient was taken ill in August with fever and unconsciousness which lasted for two days; he eventually recovered.

It is difficult, as Batten says, to locate the lesions causing the

¹ Fürbringer, P., *Deutsch. med. Woch.*, No. 3, Jan. 21, 1892.

² Batten, Fred. E., *The Lancet*, Dec. 20, 1902; and *Trans. of the Med. Soc. of Lond.*, vol. xxviii., p. 116, 1905.

symptoms. The encephalitis probably affected both the cerebrum and cerebellum as well as the sixth nerve in its course after leaving the nucleus. Whether the rhythmical spasm in this case was due to cortical motor lesions cannot be asserted. It sometimes happens, as the writer has shown in another paper on Jacksonian epilepsy,¹ that local spasm may be due to lesions at the base, as in two cases of cerebello-pontile tumour recorded by him. The probable explanation of such cases is that they are caused by irritation of the pyramidal tracts when the arm and leg are involved, or of the facial tract or the nerve itself when the face takes part in the spasm. In the case of Batten, as both cerebellar and cranial nerve symptoms were present, the rhythmical movements may have been due to involvement of the motor tracts in the pons and oblongata; on the other hand, as the lesions of polio-encephalitis are often found in several or many parts of the brain, the motor cortex or subcortex on both sides may have been the seat of circumscribed foci of encephalitis.

Spielmeyer² has contributed an article on the "Symptomatology of Cerebral Encephalitis, especially on an Epileptic Form of this Affection." This article, or a portion of it, is of considerable interest in connection with the case here recorded. According to him, either coma, paralysis, or motor excitation is always present in this affection. His group of epileptic cases, which is of particular interest here, is somewhat fully discussed. The spasms may be either Jacksonian or general, and are usually transient, but may disappear to return. I have seen several illustrations of this form of focal encephalitis in children in which the spasms, having ceased, returned, or in which they did not appear until some years after the attack of encephalitis, this having presented chiefly the features of the comatose or paralytic variety. Two personal cases are given by Spielmeyer showing marked encephalic lesion both of the cortex and subcortex of the motor region.

Of interest is the view taken by Spielmeyer that the disease which the epileptic form of encephalitis most counterfeits is brain tumour. The mode of onset and progress of the two affections

¹ Mills, C. K., *The Boston Medical and Surgical Journal*, vol. cliv., No. 17, April 26, 1906.

² Spielmeyer, W., *Centralblatt für Nervenheilkunde und Psychiatrie*, vol. xxvii., June 1904.

and the presence of optic neuritis in brain tumour furnish the best points of differentiation. The frequent occurrence of focal encephalitis in connection with infectious diseases is also a point of much diagnostic value. Attention is called to the resemblance of the symptoms of epileptic encephalitis to those of "meningitis serosa," and it is suggested that as cases of the latter affection have not been verified by necropsy, this form of so-called meningitis may be in reality an encephalitis.

Jacksonian Spasm involving the face and arm—facio-brachial monoparesis—no impairment of sensation—focus of hæmorrhagic encephalitis in the motor zone, sharply delimited posteriorly by the central fissure.

M. G., eighty-three years old, was a patient in the writer's wards of the Philadelphia General Hospital. She had been for nearly three years before her admission, October 12, 1905, an inmate of the out-wards. Her family and past history were negative, except the record of some heart disease which had existed for about a year. The illness for which she was brought to the hospital began a few days before her admission. She had three attacks of spasm involving the left side of the face and left arm. She was not unconscious during the convulsive seizure, but the left arm became weak, especially at the wrist.

Examination showed an old woman well nourished. Her tongue protruded towards the left side. The left side of the lower face was smoother than the right, and voluntary movements were not as well performed on the left as on the right. The movements of the upper half of the face were well performed on both sides, but the ocular movements were not well executed, either singly or together.

The left ocular and palpebral conjunctivæ were greatly inflamed. The pupils were moderately dilated, round, and responded well to light, accommodation, and convergence. The left upper extremity showed distinct weakness in all movements with wrist-drop. The patient could not elevate the hand nor extend the fingers. Flexion of the fingers and hands was likewise impossible, but the biceps and triceps movements and those of the muscles of the shoulder girdle were preserved to a slight degree. No paralysis was present in the lower extremity

Sensation was normal in the paralysed parts and also elsewhere throughout the body. The reflexes were normal.

During the time of examination the left side of the face and the left hand were the seat of intermittent convulsive movements. The hand was drawn somewhat to the left and the angle of the mouth was drawn outwards and upwards. The eye was closed and the brow wrinkled, but without any frowning movement or movements of the eyeballs or jaws. Dilatation of the left nostril and movement of the platysma took place. In the spasm the movement of the hand was confined to the deep extensors of the fingers and thumb and group of ulnar extensors, there being no movement of the upper arm and shoulder muscles.

At one time while examining the patient she had one of the above described convulsive attacks lasting two minutes. Consciousness was completely retained, and the movements were absent in all other parts of the body except in the arm and face as above mentioned. At the conclusion of this seizure the hand and face remained quiet for some time. These notes were made, October 1905, by Dr J. W. M'Connell, the assistant neurologist. Examination of the heart at this time showed the action of the heart to be irregularly intermittent; the pulse was small; a mitral systolic murmur was present. She was voiding very little urine.

On October 15 another examination was made, showing almost complete loss in convergence and in moving the eyes to the left and in associated upward movements, although this loss in associated movements was more marked on the right than on the left. The paralysis of the left upper extremity was deeper than when the previous note was made. No contracture was present, and the lower extremity was still not involved.

On October 16 the urine still continued to be scant, and she grew more apathetic and stuporous. The skin was clammy, and she could hardly be made to understand anything. Moist râles were distributed throughout the lungs. During the day she had six Jacksonian convulsive attacks like those already described. She died October 16, 1905.

Necropsy showed the heart to be very much enlarged, all the cavities containing post-mortem and some ante-mortem clots. The musculature of the ventricles was thickened. It was reddish pink in colour, was soft, flabby, easily pinched through,

and had a greasy surface. The mitral leaflet was thickened; the aortic leaflets were also thickened, especially the posterior. The aorta showed numerous patches of sclerosis, diffuse in character. Several ulcers were found in the abdominal aorta on its posterior wall.

The right lung was normal in size, the apex and anterior borders showing black pigmentation, in which areas the lung structure had a downy consistency. The left lung showed the same changes.

The spleen was small, and the capsule stripped easily and presented an iced-caked appearance. It cut with some resistance. The surfaces were dark red in colour and presented many islands of fibrous tissue.

The right and left suprarenal were normal.

The left kidney was normal in size, flabby, and cut with considerable resistance; the capsule was thickened and stripped with difficulty, leaving a granular surface. The cortex was narrow at points; medullary portion was purplish and congested. The pelvis contained an increased amount of fat.

The right kidney showed changes similar to those of the left. Cirrhosis of the liver was present. Examination of the stomach, intestines, pancreas, and other organs showed nothing abnormal.

On removing the calvarium, the meninges showed nothing noteworthy. A hæmorrhage was present in the right parieto-occipital fissure. A pathological area, apparently to the naked eye one of softening, was found in the precentral convolution in the arm area and upper portion of the face area (Fig. 1).

The ventricles were normal; the arteries at the base of the brain were hard and sclerotic.

A section of the pons made just cephalad of the fifth nerve showed an area possibly abnormal, this not including the motor bundles, but microscopical examinations failed to confirm this observation.

The specimens were sent to the Laboratory of Neuropathology at the University of Pennsylvania, where they were examined by Dr Wm. G. Spiller, to whom I am indebted for the following report:—

The middle third of the right precentral convolution is distinctly softer than the surrounding tissue, and on section the softening is seen to be the result of numerous hæmorrhages within



FIG. 1.—Photograph showing the area of encephalitis indicated in black, sharply limited posteriorly by the central fissure.

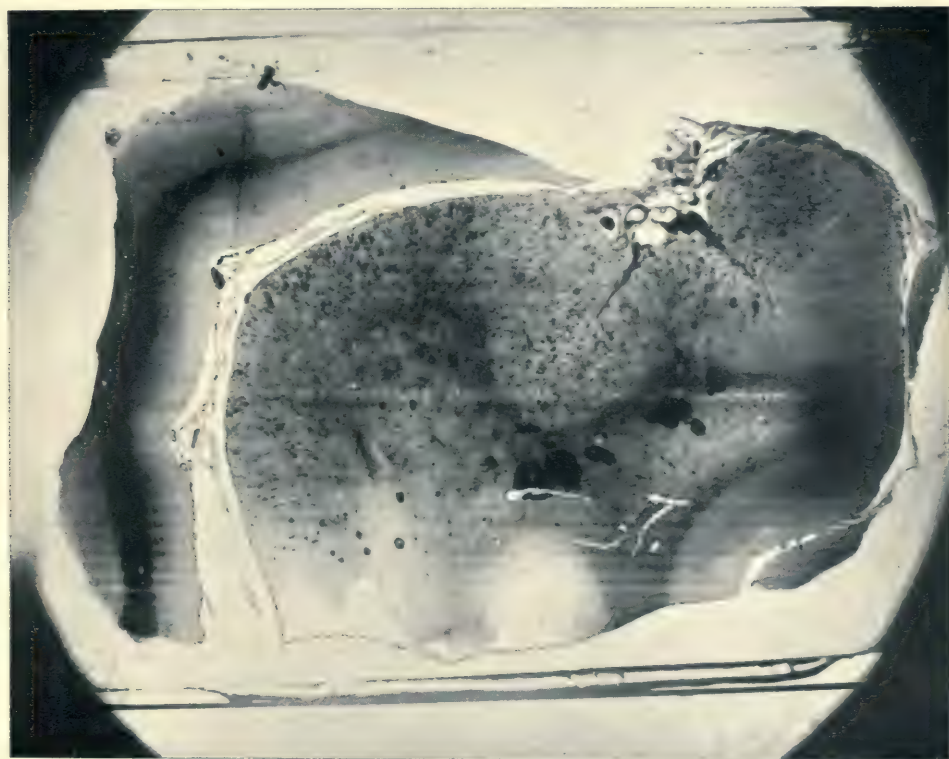


FIG. 2.—Horizontal section through the encephalitic area showing numerous minute hæmorrhages. The posterior part of the section from the postcentral convolution to the anterior portion of the precentral convolution are not implicated.

the cortex. A hæmorrhage of recent origin, about the size of a hickory nut, is in the outer portion of the right occipital lobe.

Microscopical sections through the affected precentral convolution show that the numerous minute hæmorrhages are almost confined to the cortex, although a few are in the subjacent white matter. At the region of greatest extension the anterior one-fifth of the precentral convolution alone escapes. The hæmorrhages extend deep into the central fissure, but do not pass beyond the bottom of the fissure into the post-central gyrus. The diseased cortex stands out very sharply to the naked eye. The transition from the normal to the hæmorrhagic cortex is quite sharp, even under the microscope. The pia adjacent to the affected cortex is also hæmorrhagic. A considerable amount of black granular pigment is found in the hæmorrhagic cortex. Numerous fatty granular cells and round cells are in the pia over the hæmorrhagic cortex. A small round hæmorrhage about .5 cm. in diameter is found at the extreme lower end of the hæmorrhagic area. The sharp limitation to the precentral gyrus, and the almost complete limitation to the cortex of this gyrus and to its posterior half are very striking. There is no round-cell infiltration about the chiasm, but there is slight round-cell infiltration about the small vessels of the cerebral peduncles (Fig. 2).

The symptomatology and pathology of this case speak for themselves in the light of the short study of the literature of the subject prefixed to the history. The strict limitation of the lesion to the cortex anterior to the bottom of the central fissure is of interest in connection with the question of the absolute precentral position of the motor zone. The disturbances of ocular movements and the hemianopsia apparently present were probably due to the gross hæmorrhage in the occipital region.

MYOPATHY OF THE DISTAL TYPE AND ITS RELATION TO
THE NEURAL FORM OF MUSCULAR ATROPHY (CHAR-
COT-MARIE, TOOTH TYPE).*

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FROM THE DEPARTMENT OF NEUROLOGY AND THE LABORATORY OF NEURO-
PATHOLOGY IN THE UNIVERSITY OF PENNSYLVANIA, AND FROM THE
PHILADELPHIA GENERAL HOSPITAL.

Various forms of muscular atrophy have been distinguished and sharply separated from one another. While it is possible to make these classifications for typical cases, the borderline cases cause great confusion, and make clinical diagnosis at times extremely difficult.

The primary neurotic muscular atrophy of Charcot-Marie and Tooth was supposed to be so sharply defined that confusion with other types of muscular atrophy could not occur. The characteristics of the type as given by Charcot and Marie¹ are:

Progressive muscular atrophy implicating first the feet and legs, and not appearing in the upper limbs (hands and later forearms) until several years later; the progression of the atrophy being slow.

Relative integrity of the muscles of the limbs near the trunk, or at least, much longer preservation of these than of the muscles of the distal ends of the limbs. Integrity of the muscles of the trunk, shoulders and face.

Fibrillary contractions in the atrophying muscles.

Vasomotor disturbances in the portions of the limbs atrophied.

No pronounced contractions of tendons in the atrophied limbs.

Sensation usually intact, but sometimes affected.

Cramps frequent.

Reaction of degeneration in the atrophying muscles.

Commencement of the affection usually in childhood, the disease often found in several brothers and sisters, and sometimes in the previous generations.

*Read before the American Neurological Association, June 4 and 5, 1906.

¹Charcot and Marie. *Revue de médecine*, 1886, p. 97.

Charcot and Marie based their conclusions on five cases of their own, and on a few cases in the literature which they believed belong to this type.

I think it well to emphasize the fact that Charcot and Marie in their original paper acknowledged the possibility of implication of the muscles of the limbs near the trunk, at least to some extent. They said that the muscles of the thighs seem to preserve their power and volume during a certain period, but that this integrity often is not real. The vastus internus is first involved. In their summary at the end of their paper they speak of relative integrity of the muscles of the limbs near the trunk, or at least, much longer preservation of these muscles than of those of the distal ends of the limbs. Unquestionably, however, they emphasized the earlier and greater involvement of the muscles at the distal ends of the limbs as the most characteristic feature of the disease.

If we insist on one of the features emphasized by Charcot and Marie; viz., the almost constant absence of all contraction of tendons, many cases classed as progressive neurotic muscular atrophy must be regarded as doubtful.

Cases conforming strictly to the type as described by Charcot and Marie occur, and these probably have a distinct pathology. Such a case is the following, which has been in the Philadelphia General Hospital many years, several times under my care, and so far as I know has not been reported:

N. Hutt, 59 years of age, hostler, was admitted Feb. 15, 1901, at which time the following notes were made:

Chief complaint: Weakness in the arms and legs.

Family history: Father died at about the age of 60 years from some affection of throat, with which he had been ill for one month. Mother died at about the age of 60 years from a second "stroke" of apoplexy. One brother died in infancy and four brothers and three sisters are healthy. The patient is not aware of any family diseases.

He has had only the milder diseases of childhood, and was healthy as a boy and a man, except for the present complaint, and typhoid fever at 17 years. He says he was a moderate user of alcohol, positively denies venereal disease, and has never been married.

History of present illness: He states that at the age of 15 years he began to have pains in the legs and feet, which would last for a week or two, and incapacitate him for work.

These attacks seem to have come on usually in the spring of the year, and he would then be free until the next spring. He had no weakness of the limbs, and was able to work as a hostler, though at about the age of 35 he began to have pain in his hands and arms, and he noticed that he was becoming weak in



Fig. 1. The neurotic form of muscular atrophy, hands and forearms involved. (Photographed by Dr. Ralph Pemberton.)

the legs. These grew thinner and became somewhat deformed. From that time until the present the progress of the disease has been constant, though he has now much less pain than formerly, and only in his feet. During the past five years he has been unable to do any work. The atrophy of the muscles of the

legs preceded that of the upper limbs ten years, according to the patient's statement.

He is well nourished, pupils are equal and react to light. Tongue is clean and shows no tremor. Chest and abdomen are well formed, the expansion of the chest is fair, lungs are normal, heart sounds are muffled and distant.



Fig. 2. The neurotic form of muscular atrophy, hands and forearms involved. (Photographed by Dr. Ralph Pemberton.)

Examination of eyes by Dr. Shumway, Oct. 20, 1904: The pupils are equal, respond promptly; eye movements are good.

Ophthalmoscopic examination: Haziness of lens, retinal veins full and show pressure signs, result of arteriosclerosis,

otherwise fundus is normal. Vision, O. D. 5-8; O. S., 5-10; fields are normal.

Condition of the patient at my examination March 7, 1906: The atrophy is intense in the hands (figs. 1 and 2), distinct in the forearms, but the arms above the elbows are well developed. Sensations for touch, pain and temperature are normal in the



Fig. 3. The neurotic form of muscular atrophy, feet and legs (not thighs) involved. (Photographed by Dr. Ralph Pemberton.)

upper limbs. Sense of position is normal in the fingers. The grasp of each hand is very feeble. The muscles of the trunk and head are well developed. He raises the upper limbs above the head with much power, and is able to dress himself and handle his clothing with considerable skill, notwithstanding

the intense atrophy of the hands. Biceps and triceps reflexes are weak.

The lower limbs, above the knees, are well developed, and if there is any atrophy here it is very slight. Below the knees the atrophy is marked. The feet are inverted and talipes equinovarus (fig. 3) is very pronounced on each side. The patellar reflex and Achilles tendon reflex are lost on each side. The toes are somewhat cyanotic. Sensations for pain, touch and temperature are normal in the lower limbs. He is able to walk without assistance and without crutch or cane, but his gait is rather slow, with legs far apart and his feet turned inward. His gait is somewhat steppage in character.

If we hold to the type of muscular atrophy as presented by this patient; viz., atrophy confined strictly to the distal parts of the limbs, we shall be able to recognize the neurotic muscular atrophy as a distinct type, with a distinct pathology. The cases of this kind with necropsy are very rare, we are obliged to depend on the findings of Marinesco, Sainton, and Dejerine and Armand-Delille. Oppenheim includes the cases of Dubreuilh and Siemerling, but Sainton regards the former as far from typical, and does not refer to the latter, which should, I think, be excluded.

In Siemerling's² case there was great atrophy of all the extremities in the upper parts as well as in the lower, and of the trunk; flaccid complete paralysis of the lower limbs, so that all voluntary movements of these limbs were lost. Siemerling found degeneration of the posterior and lateral columns, most intense in the lower thoracic and upper lumbar regions, degeneration of peripheral nerves and muscles; atrophy of the cells of the anterior horns, of the columns of Clarke, of the anterior roots, and of the spinal ganglia.

He thinks there can be no doubt that the case belongs to the spinal neurotic atrophy, but if we accept this opinion we cannot limit the type to those cases in which the atrophy affects the distal portions of the limbs, and the proximal portions and trunk escape.

In the case of the neurotic muscular atrophy studied by Paul Sainton³ the lesions were:

Sclerosis of the posterior columns, especially of the columns of Burdach. Slight degeneration of both pyramidal tracts. Al-

²Siemerling *Archiv. für Psychiatrie*, Vol. 31, 1898, p. 105.

³Paul Sainton *Nouvelle Iconographie de la Salpêtrière*, Vol. 12, 1899.

teration of the columns of Clarke. Atrophy of the cells of the anterior horns. Slight degeneration of the intramuscular nerves. Slight sclerosis of the nerves of the forearms and legs, very distinct only in the peroneal nerves. Atrophy of the muscle fibers even causing complete disappearance of some fibers with proliferation of connective tissue.

In this case the atrophy began in the upper limbs. The lesions resembled those observed by Marinesco in his case, except that Marinesco found the anterolateral columns intact. Judging from these two cases Sainton regards as the lesions of this form of muscular atrophy: sclerosis of the columns of Burdach and Goll, atrophy of the cells of the anterior horns, and alterations of the peripheral nerves of varying intensity, sometimes slight.

The lesions found by Dejerine and Armand-Delille⁴ in a case of neurotic muscular atrophy were degeneration of the posterior columns, degeneration of some of the nerve cells of the anterior horns of the cervical and lumbar regions without diminution in the number, chronic meningitis, degeneration of the muscles of the hands and feet, alteration of the intramuscular nerve fibers in the hands and feet (*i.e.*, many nerve fibers of small size, many empty nerve sheaths and a few nerve fibers in the process of degeneration). The nerve trunks, the cutaneous sensory nerves, and the anterior and posterior nerve roots with slight exception were normal.

The literature contains many cases reported as belonging to neurotic muscular atrophy in which some atypical features were present. Sachs⁵ two cases were probably the first reported in America. In his first case the patient had pronounced kyphosis of the lumbo-sacral region, slight wasting of the infraspinatus muscle, and decided diminution in the power of thighs as well as of legs.

The younger brother of the first patient had general emaciation of all parts of the upper extremities, very distinct atrophy of the infraspinatus and marked general atrophy of the legs, which seems to imply involvement of the thighs.

In discussing the cases together Sachs says the atrophy caused a weakness of the thigh muscles as well as of the muscles

⁴Dejerine and Armand-Delille. *Revue Neurologique*, 1903, p. 1198.

⁵Sachs, *Brain*, 1890, Vol. 12, p. 447.

of the legs, and he speaks of atrophy involving all the muscles of the lower extremities as common in the neurotic muscular atrophy.

In one of Hoffmann's⁶ cases the lower limbs were wasted in all parts, and chiefly below the knees, but the strength of the thigh muscles was good. Fibrillary tremors were not observed. In another case, Hoffmann found wasting of the upper part of the left trapezius muscle and flattening of the supraspinous and infraspinous fossæ, without any loss of function. Fibrillary tremors were not seen. In another case he says the thigh muscles were distinctly weak and much wasted, especially in the lower part of the thigh. The thoracic muscles were poorly developed and the ribs were prominent. In still another case all the thigh muscles were much atrophied, as well as the muscles of the buttocks and the lower part of the extensors of the back. The patient had a distinct lumbosacral kyphosis, and experienced difficulty in rising from the ground, such as is seen in progressive muscular dystrophy. The legs were kept far apart and the patient climbed upon himself as in muscular dystrophy.

Hoffmann accepts involvement of the facial muscles as a part of the neurotic atrophy.

In the case reported by George W. Jacoby,⁷ and recorded as one of progressive muscular atrophy of the peroneal type, severe pain was felt in the lower extremities when the child was two years old. When she was four years old one leg was found to be weaker and thinner than the other. Later atrophy of the thigh on the side opposite to that of the affected leg was noticed. There was marked lordosis and slight lateral curvature. The muscles of both thighs were in a continual state of unrest, almost like fibrillary twitchings. The right foot was in equinovarus position. There were no sensory disturbances, but there was partial reaction of degeneration in the affected muscles. The arms were not affected.

Jacoby acknowledged that the case was not typical, inasmuch as the atrophy, although bilateral, was asymmetrical, one leg being affected and the opposite thigh.

⁶Hoffmann. *Deutsche Zeitschrift für Nervenheilkunde*, Vol. 1, p. 95.

⁷G. W. Jacoby, *JOURNAL OF NERVOUS AND MENTAL DISEASE*, 1894, p. 259.

Sachs, however, concurred in the diagnosis of muscular atrophy of the peroneal type.

In at least one of Diller's⁸ two cases (brother and sister), the thighs also were wasted, and the patient climbed upon his lower limbs as is seen in muscular dystrophy. He had left talipes equinus. Response to electricity in the paralyzed legs was very sluggish and reaction of degeneration was obtained.

In the first of the two cases reported by Given Campbell⁹ the disease progressed in the lower limbs until these limbs became useless and the muscles of the thighs also were much wasted. The deltoid muscles, especially the left were weak. The weakness and wasting developed hand in hand, and began in the peroneal group of muscles. The small muscles of the hands were not affected. The muscles of the trunk were not wasted, unless possibly the abdominal muscles to a slight extent.

I could add to these cases others to show that atypical features have not prevented the diagnosis of muscular atrophy of the Charcot-Marie-Tooth type, and therefore it is not surprising that cases of muscular dystrophy have been diagnosed clinically as belonging to the type. In this connection it is interesting to read Sachs' words published in 1889 (Brain) when the peroneal type of muscular atrophy was little known. He said: "There is a consensus of opinion, however, on this one point, that the cases in question do not belong to the category of primary muscular dystrophies." He was speaking of the peroneal type of atrophy. Oppenheim and Cassirer¹⁰ showed that this consensus of opinion no longer exists.

They pointed out that the clinical picture of the neurotic muscular atrophy is not so sharp as Hoffmann believed, and that the pathology is still uncertain. In their case the muscular atrophy had existed about two years. The symptoms began with severe pain in the lower and upper limbs. The distal portions of the lower limbs, especially those innervated by the peroneal nerves, were affected, and in the upper limbs only a part of the small hand muscles, and later the triceps and supinator longus, were implicated. In some muscles incomplete reaction of degeneration was obtained, in others all electrical response

⁸Diller. Philadelphia Medical Journal, March 17, 1900, p. 642.

⁹Campbell. JOURNAL OF NERVOUS AND MENTAL DISEASE, 1900, p. 274.

¹⁰Oppenheim and Cassirer. Deutsche Zeitschrift für Nervenheilkunde. Vol. 11, p. 143.

failed. The tendon reflexes were diminished. The orbicularis palpebrarum on each side was involved. Later the calf muscles became atrophied. The diagnosis seemed to lie between chronic multiple neuritis and neurotic muscular atrophy. The central and peripheral nervous systems were normal, but the muscles were much degenerated.

As regards the involvement of the orbicularis palpebrarum, Oppenheim and Cassirer refer to the fact that in Sachs' cases the infraspinati were atrophied, in a case of Hoffmann's the upper portion of the left trapezius and the supraspinati and infraspinati were atrophied, in a case of Donkin's reaction of degeneration was obtained in the left trapezius, in a case of Hülemann's the pectoral, rhomboid, latissimus dorsi muscles were somewhat atrophied, in a case of Eisenlohr's the atrophy of the upper limbs was like that of myopathy, but of the lower limbs like that of the neurotic muscular atrophy. All these cases were regarded as belonging to neurotic muscular atrophy, and yet they showed some features in common with muscular dystrophy. Especially interesting are two cases in sisters observed by Dähnhardt, in one, the wasting resembled the neurotic muscular atrophy, in the other the progressive muscular dystrophy.

Oppenheim and Cassirer concluded that the clinical picture of progressive neurotic muscular atrophy may be caused by muscular dystrophy, and that the pathology of the former is not always the same and not as yet clearly defined.

Gowers¹¹, in reporting a clinical case of distal myopathy in 1902, said he had not seen a similar case nor did he know that one had been recorded. The patient, a boy, was eighteen years old, and was the only member of the family affected. The symptoms first attracted notice when he was ten or twelve years old; then it was noticed that he often caught his toes against the ground in walking. At a later date his hands were found to be weak. The feebleness of hands and feet steadily increased until it became pronounced. He was unable to flex the ankles, although he could just extend the toes and could move each foot slightly in and out by the tibialis anticus and the peronei. He could extend the ankle joint by the calf muscles with some force. The movements of the knee and hip were performed with good power, the knee jerks were present, but the left was less than

¹¹Gowers. *British Medical Journal*, July 12, 1902.

the right. The thigh muscles were of normal size; the anterior tibial muscles were distinctly smaller on the left side; the calves were rather large and firm, and were sufficiently large to suggest a resemblance to pseudo-hypertrophic paralysis. The grasp was extremely feeble, with the right hand he could not move the dynamometer, and with the left he could only move the index to 2 k° instead of 50 or 60. Extension was also feeble; he could get the fingers with the wrist into line with the forearm, but could not fully extend them when the wrist was overextended. The muscles of the forearms and hands were small, but presented no wasting comparable to that which is seen in progressive spinal atrophy. Above the forearm the muscles had fair power and presented no wasting; only a trifling atrophy of the middle part of the trapezii could be observed. Electrical excitability was lowered in the affected muscles in proportion to their feebleness, and equally to faradism and voltaism.

The neck muscles were normal except the sterno-mastoids. He could not raise the eyebrows at all, and closure of the eyes by the orbicularis muscles were weaker than normal.

Gowers said this case differed from all recognized forms in the purely distal distribution of the affection in the limbs and the normal state of the muscles near the trunk. The case resembled therefore the neurotic muscular atrophy, but the implication of the sterno-mastoid, frontalis and orbicularis palpebrarum muscles probably prevented Gowers from classing the case under this head.

In the case of myopathy of the Aran Duchenne type reported by Dejerine and Thomas,¹² the muscular atrophy was confined to the upper limbs and the hands were much wasted. The muscles of the head, neck, face, trunk and lower limbs were not at all affected. Fibrillary contractions were seen in the upper limbs. The symptoms began when the patient was 49 years old, and lasted thirty-one years. It is not surprising therefore that the case was regarded as one of myelopathy during the lifetime of the patient. The spinal cord and nerves, even the intramuscular nerves, were normal. The muscles were much altered, many muscular fibers had disappeared, and those that remained were much atrophied, many had lost the transverse

¹²Dejerine and Thomas. *Revue Neurologique*, No. 24, Dec. 30, 1904.

and longitudinal striations, the sarcolemma nuclei were multiplied and some were enlarged.

The case of muscular dystrophy affecting the hands and feet reported by C. Macfie Campbell¹³ was without necropsy. Other members of the family were similarly affected. Fibrillary twitching in the interossei and short flexor of the thumb, and reaction of degeneration in two of the atrophied muscles of the lower extremities raises the suspicion that the case possibly may belong to the type of neurotic muscular atrophy.

These are the only instances in literature I know of where muscular dystrophy gave distinctly the clinical signs of neurotic muscular atrophy, but a case of this character with necropsy has come under my observation. I hesitated during the lifetime of the patient to make the diagnosis of neurotic atrophy, although the case was regarded by some as belonging to this type. I hesitated because the thighs and upper arms and trunk were also much atrophied, and I felt that the diagnosis should be left open. The commencement of the atrophy in the lower part of the lower limbs, the slight reaction of degeneration in these parts, the talipes equino-varus on each side, the symmetry and the later involvement of the upper limbs suggested the neurotic form. The microscopical examination has shown a normal central and peripheral nervous system with much atrophy and disappearance of muscle fibers.

G. R., age 28 years, was admitted to the Philadelphia General Hospital Oct. 11, 1898, complaining of pain about the knee and difficulty in walking.

Family history: Father died of heart disease at about 25 to 30 years of age, mother living and well, at 50 years of age.

The patient was married at 23 years of age. He has one child, three years old, in good health. He uses neither tobacco nor alcoholic drinks, and has been employed as a watch case engraver since his fifteenth year.

Past medical history: He has had childhood diseases. When ten years old he fell and injured his back and could not walk for two months (the condition was diagnosed as spinal meningitis), but he recovered the use of his limbs satisfactorily in about one month after getting up. Three years ago he had an attack of cystitis.

History of present illness: About two years ago he fell from a street car, striking on his hip and back of his head,

¹³Campbell. Review of Neurology and Psychiatry, March, 1906, p. 192.

badly bruising his right hip and thigh. He suffered no particular inconvenience from this at the time, except a slight but constant pain in the knee joint. About six months later, upon arising one morning, he fell to the floor and found himself unable to walk, but this was probably not the first manifestation of weakness. Two or three days later he was able to walk with a cane, and after a few days could walk without it. He walked without a cane about two weeks. During this time, while walking along the street, he fell and had to be assisted to a car, and could not walk during the remainder of the day. About a week after this he was obliged to use the cane again, and has continued doing so since. For the past year and a half the inability to walk has steadily progressed, and coincident with this has been the muscular wasting. No involvement of bladder has occurred, except a cystitis three years ago, previous to his accident. He had noticed weakness first in the left foot, later in the right foot, and three months still later his upper limbs became affected. He stated that the wasting began in the feet, extended to the legs and thighs, and that the upper limbs were affected first in the distal parts.

Condition upon admission: He is a somewhat poorly nourished adult male, of above average height; pupils are dilated, the left more so than the right, both react to light, accommodation and convergence. Speech is normal. Tongue protrudes straight and shows no tremor. Pulse is small, quite irregular; arteries are soft.

Chest and lungs: Chest is somewhat emaciated; expansion is good. Lungs are normal.

Heart: Apex beat is visible and palpable in the 6th interspace, $1\frac{1}{2}$ inches within the nipple line. First sound is loud and strong; second sounds are both accentuated. Liver and spleen are normal.

Arms: Grip, right hand, is 21; left is 17. The arms are emaciated, but not atrophied. Ataxia is not detected.

Legs: Marked general wasting of the muscles, apparently equal in both legs. Foot-drop is marked. Toes are slightly flexed. Spasticity of legs is not present.

Reflexes: Knee jerks are absent; no ankle or patellar clonus; superficial reflexes are present. Tactile sensation is preserved and normal. Pain and temperature sensations are normal.

Nov. 7, 1898: Patient's physical condition is improved since admission. No change in walking, or in rising from chair. He is gaining slightly in weight, but complains of some pain in the legs and ankles.

An examination of the eyes April 14, 1899, by Dr. Charles A. Oliver showed healthy eyegrounds; paresis of left external rectus and left superior oblique, with diplopia, which had been present a few days. This was not persistent.

At this time he was unable to walk, and could not stand alone,

although he could move his legs somewhat. Foot drop was marked on each side. The legs could not be extended. Muscular atrophy had been progressive.

On June 25, 1899, a note was made that the patient could walk, but had inversion of the foot on attempting to walk.

Sept. 10, 1899, the patient was granted a two-days pass on which he went out Friday. He was brought back by a patrol wagon last night, having been picked up at Market St. ferry, where he had suddenly lost all power of his legs. When admitted he was complaining of abdominal and lumbar pain, loss of power in legs as well as a feeling of complete exhaustion. This evening he is much better, and already can move his legs fairly well, has lost much of the lumbar and abdominal pain, and feeling of exhaustion.

Sept. 26, 1899. He has improved rapidly since his return; he can walk, but is still kept in bed.

April 13, 1901. Examination by Dr. Pearce. Both upper extremities, both above and below the elbow, respond quickly and well to faradism. There is no reaction of degeneration. Elbow jerks and biceps jerks are present, but much diminished. There is no localized wasting in upper extremities. In the lower extremities there is marked quantitative change below the knees, and very slight qualitative change in the anterior and posterior groups of muscles. There is no sensory change anywhere. Knee jerks are absent. Plantar reflex is present. No arthropathies; pupillary reactions are normal.

July 19, 1903. He has not walked for two and a half years. Sensations of touch and pain are normal.

Aug. 20, 1903. Dictated by Dr. Weisenburg. Patient is unable to walk; he can flex the right thigh on the hip slightly; left thigh movement is very poor. No movements possible in ankles; he can flex or extend the toes of both feet very slightly. Both feet are in a position of talipes equino-varus, the left more than the right. Both feet are cold. Movements of both upper limbs are free in all directions. Grip in the hands is almost nil. No power in elbows at all. Power in shoulders fairly good. Both upper extremities, shoulders, back and chest are wasted; abdomen not so much. Wasting is about equal on both sides. Thenar and hypothenar eminences on both hands are almost absent. Atrophy of interossei is marked. In the forearm the flexor group is more atrophied than the extensor; the biceps is atrophied on each side. Deltoid is slightly atrophied. Pectoral muscles wasted, also the interossei on both sides. Supraclavicular and infraclavicular fossae well marked, also interscapular region. All muscles of the shoulder girdle are atrophied, including the latissimus dorsi on each side. No fibrillary twitching is noticed in the muscles. All reflexes in the upper limbs are lost.

He has never had any urinary or rectal disturbances. The pupils are equal, respond to light, accommodation and convergence. No apparent weakness of left internal rectus or left superior oblique. No involvement of 5th or 7th nerves. Tongue protruded straight; no tremor or atrophy. No atrophy of the muscles of the face or neck.

At present the wasting in both lower extremities is about equal. The femoral group on each side is very much wasted;

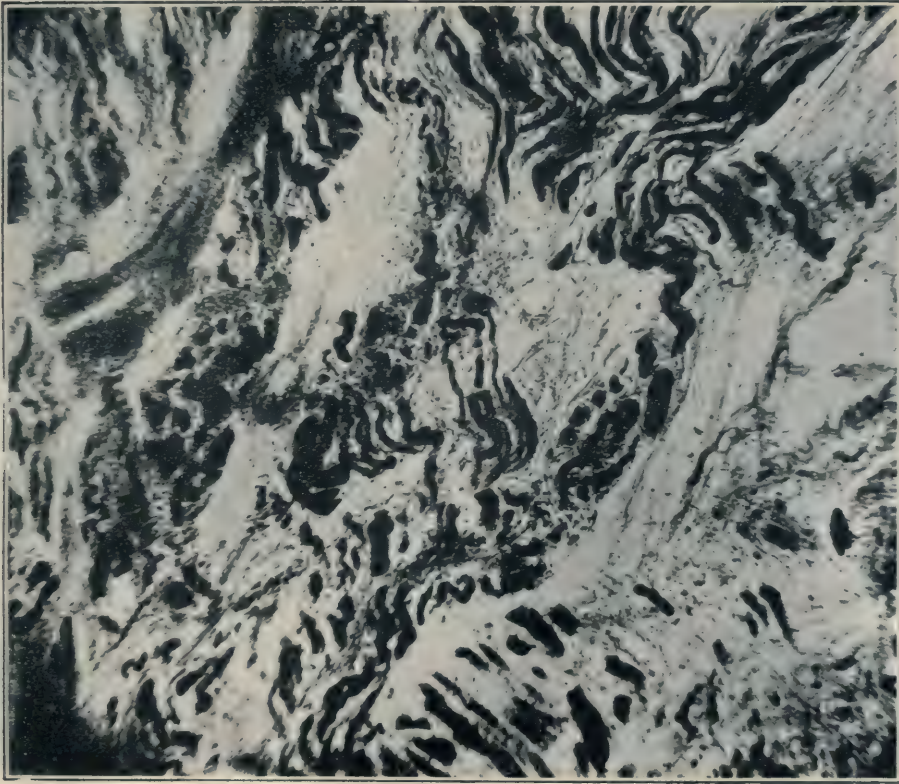


Fig. 4. Piece of muscle from the sole of the foot in a case of myopathy of the distal type. (Photographed by Dr. Alfred Reginald Allen.)

the calf muscles are wasted, as are the thigh muscles of each side. All the reflexes in the lower extremities are lost. Plantar irritation produces flexion of all the toes. Abdominal and cremasteric reflexes are present and active.

He says he has dull pains in the legs and arms, but has never had any sharp shooting pains, nor any sensory disturbances like pins and needles. Sensation is apparently normal.

Oct. 12, 1904. Dr. Shumway: Right vision, 5-5; left vision, 5-6. External ocular movements are normal. Pupil reaction is prompt. No diplopia in any part of the field. Fields full; perfectly normal eyegrounds.

July 21, 1905. Examination by Dr. Pickett: Ankles are almost fixed in position of foot drop. The right knee cannot be extended completely. Strongest movements of the body are extension of the feet (plantar flexion), supination of the fore-

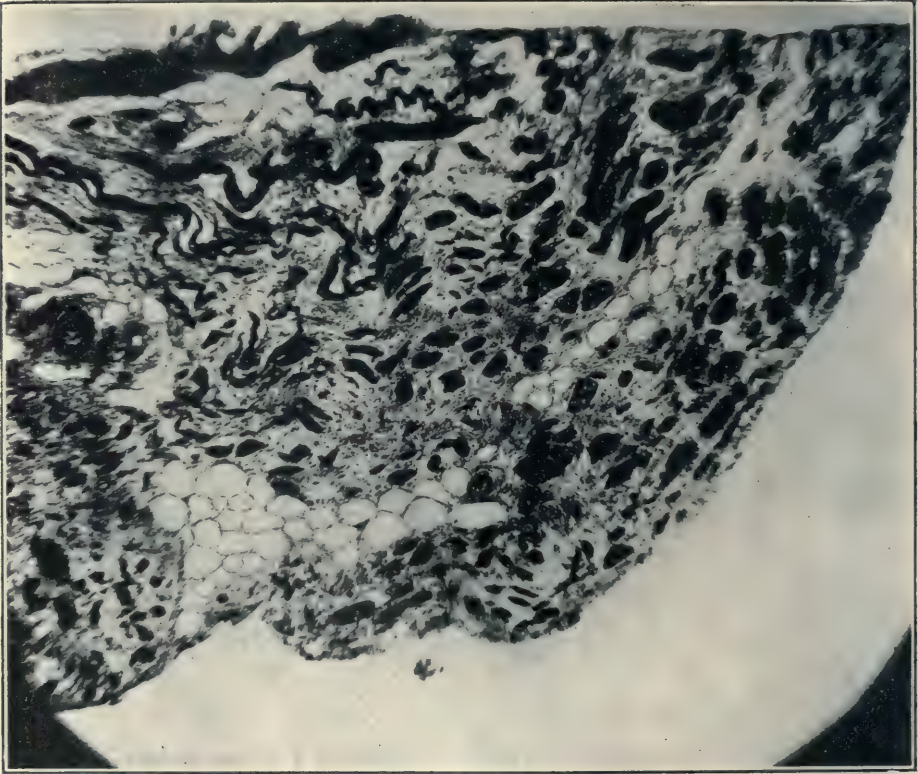


Fig. 5. A piece of muscle from the sole of the foot in a case of myopathy of the distal type. (Photographed by Dr. Alfred Reginald Allen.)

arm, then after this pronation and extension of wrist and fingers. Preserved supinators and extensors make the forearm bulge as if it and the bones were bent. His movements in dressing are at times so irregular by the unequal strength of the different muscles as to constitute a kind of ataxia.

Dec. 6, 1905. Patient's nervous condition is the same as described above. He has a cough and expectorates moderately;

has night sweats, and the sputum shows the presence of tubercle bacilli.

Urine analysis: Dec. 6, 1905. Yellowish; slight sediment: acid reaction; Sp. Gr. 1010. No sugar or albumin. Microscopic: red blood cells, a few; few calcium oxylates, epithelial cells and debris.

He died Jan. 11, 1906.

This man was examined frequently and lectured upon by me several times. He was repeatedly in my charge at the hospital. His condition at the time of death was as follows:

The lower and upper limbs were greatly atrophied and equally so at all parts. Contracture at the right knee was present, so that the leg could not be fully extended upon the thigh. The left leg could be fully extended on the thigh. There seemed to be contracture at the right hip. Talipes equino-varus was present on each side. No contractures were observed in the upper limbs. Emaciation was shown also in the muscles of the trunk and face. Slight lordosis was present in the lumbo-sacral region, and there was some protrusion of abdomen, probably secondary to the lordosis. There was no evidence at the necropsy of old spinal injury, and excepting the lordosis, there was no deformity of the vertebræ. The wasting of the face may have been caused by tuberculosis, as it was not present in 1903.

Sections from the cervical and lumbar regions of the spinal cord stained by the hematoxylin method of Weigert or the method of Marchi, by acid fuchsine, hemalum or thionin are normal. A piece of muscle from the foot (fig. 4 and 5) shows no recent degeneration by the Marchi method, but the long-standing degeneration is pronounced, and is better shown by other methods of staining. The fibrous and fatty connective tissues are much increased, the muscle fibers are greatly atrophied. The longitudinal and transverse striations are well preserved, and the sarcolemma nuclei are increased in number in some of the muscle fibers. Nerve bundles between the muscle fibers stain well by the Weigert hematoxylin stain, as do also sections of one of the plantar nerves. No degeneration is seen by the Marchi method in the latter, but the blood vessels of the nerve are thickened. A piece of muscle from the back of the trunk appears normal.

Psychasthenic Attacks Simulating Epilepsy

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PSYCHASTHENIC ATTACKS SIMULATING EPILEPSY¹

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University of Pennsylvania; Neurologist to the Philadelphia
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We are occasionally brought in contact with persons who give a history of attacks that on superficial examination might be regarded as epileptic, and yet it is questionable whether they should be classed either under epilepsy or hysteria. Recently a boy, five years of age, of Jewish parentage, was brought to me with a history of peculiar attacks. He had had these since teething. He had not had any aura, had never bitten his tongue nor voided urine nor injured himself in an attack, but he falls to the ground, does not attempt to catch hold of any support while falling, lies quietly without any convulsive movements, is unconscious about five minutes, and after the attack is drowsy about half an hour. I have been unable to find hysterical stigmata in the little patient, but the examination was difficult. His father informed me that the boy never had an attack unless his feelings had been hurt. Two children in the family, a little older than the patient, have had attacks of exactly the same character; in one child they lasted until the age of two years; in the other not quite so long. It is difficult to make a diagnosis in such cases as these.

In the *Journal fur Psychologie und Neurologie*, Vol. VI., 1905-1906, Oppenheim discusses peculiar attacks under the title of Psychasthenic Convulsions. Although he has spoken briefly of this condition previously, he gives in this recent paper a full presentation of his views. Convulsions may occur in certain forms of neurasthenia, in cases where there is not hysteria, nor epilepsy, nor organic disease. The first contribution to this subject was made by Westphal in 1872

¹Read at the joint meeting of the Philadelphia and New York Neurological Societies, held in Philadelphia, November 24, 1906.

(*Archiv fur Psychiatrie*, Vol. III.) in his paper on agoraphobia, and according to this author the occurrence of convulsions with agoraphobia is not uncommon, and they may be seen as frequent signs of various psychopathic and neuropathic conditions. Oppenheim refers to the fact that Westphal's views have not met with general acceptance.

As Oppenheim presents the subject, the individuals are intensely neurotic or psychopathic from birth, and show the first symptoms of this diathesis in childhood. The neurasthenia is of the grave type which has been regarded by French writers, especially Janet and Raymond, on account of mental abnormalities, as psychasthenia. The tics, states of anxiety, phobias, obsessions and vasomotor disturbances predominate. On such a foundation, with, however, some immediate cause such as emotional disturbance, mental or physical overwork, alcoholic indulgence especially by one unaccustomed to it, sleeplessness or a period of anxiety the attack may develop.

This may be only deep unconsciousness with involuntary defecation and micturition, or there may be also convulsions, biting of the tongue and rigidity of the pupils. Usually only a few of these attacks occur, interspersed with periods of vertigo, anxiety, etc., and the tendency may disappear under proper hygienic treatment.

These attacks are not hysterical, every hysterical stigma is wanting, and the attacks themselves are not hysterical in character. Oppenheim dismisses the question of any resemblance to hysteria in a few lines. It would be better had he devoted more space to this aspect of the subject. The differentiation from epilepsy is more difficult :

(1) The attack in itself cannot be distinguished from that occurring in epilepsy. The patient is not an epileptic, he has not had such attacks in childhood or early youth, he is always neurasthenic or psychasthenic, and always periods of anxiety, phobias, tics or vasomotor disturbances have preceded the convulsions.

(2) A special cause for the convulsive attack is always necessary, such as over-exertion, mental or physical anxiety, vertigo, etc.

(3) The attacks are merely episodes in the course of the psychasthenia; they may be few or there may be only

one during the life of the individual.

(4) The attacks may resemble fully the epileptic, but on the other hand there may be variations, thus profound unconsciousness may occur without convulsions, or the convulsions may be limited to a few muscles, or they may persist after consciousness has returned. They may resemble *petit mal*.

(5) Intelligence and memory do not become impaired even though the attacks may be numerous.

(6) Treatment should be mental, bromides are of little value.

Acquired neurasthenia probably never causes these convulsions. Oppenheim prefers the name of "psychasthenic convulsions," even though convulsions are not always present; "psychasthenic attacks" he regards as too comprehensive, but it seems to me that the latter designation is preferable. Mistakes of diagnosis may be made easily, indeed Oppenheim himself has made them, as in one of his cases an organic condition was present. I am inclined to think that the danger of mistake is especially great as regards the dreamy state of epilepsy described by Hughlings Jackson under the name of "uncinate group of fits."

Attacks of ambulatory automatism may belong to a psychopathic diathesis. In one of Oppenheim's cases, pain which he regarded as neurasthenic or hysterical was present. His first case resembles closely one of the two that I report.

A young woman, when in a crowd, as in the theatre or on the street, became possessed with the idea that she saw a person bleeding from the nose, she passed into a condition of anxiety or fear, sometimes became unconscious, fell, and voided urine and feces. The description of this case is very brief, and it is not stated whether the person seen was always the same or not.

I have not been able to find any reply to Oppenheim's views, and as yet they seem to have received little attention. The subject is, however, one of importance, because of the resemblance of these conditions to epilepsy, and it is most desirable to make the differential diagnosis. The word epilepsy conveys much dread to the patient and his relatives, but far more important is the fact that not only the diag-

nosis, but also the treatment and prognosis of the psychasthenic attacks are essentially different. These psychasthenic attacks are very different from those known as psycholeptic crises.

In these, as described by Janet, we have a very different condition. The attack may come on suddenly, and terminate suddenly, or in another form may terminate imperceptibly, after running an almost indefinitely prolonged course. The chief feature of the attack is the sense of unreality, both for the person himself and his surroundings, but there is no mental confusion, the memory may even be more active, there are no convulsions and no disturbance of voluntary movements. It is a dream-like state.

Janet¹ also shows that there are certain features in common between psychasthenia and epilepsy, that an epileptic may be a psychasthenic, but this is not the same as saying that a psychasthenic may have attacks exactly like those of epilepsy and yet not be epileptic. It is very probable that many will not dismiss the resemblance of these psychasthenic attacks to hysteria so briefly as does Oppenheim.

The question largely depends on what we shall call hysteria; for example, Morton Prince² regards as signs of, but not peculiar to, hysteria fixed ideas and obsessions, and most frequently of all the neurasthenic state. Hysteria, according to the recent presentation of the subject by Babinski³ and a few others is a peculiar mental condition in which the disturbances may be produced by suggestion and cured by persuasion, while Janet⁴ would omit the latter part of the definition having reference to cure by persuasion, but he includes obsessions, phobias, etc., under his designation of psychasthenia.

There are several recent papers in which the resemblance of hysteria to epilepsy is discussed. Unquestionably many of the psychoneuroses are related, and the boundary lines cannot be sharply drawn, but that does not imply that they should be abolished. We are not yet at that point where

1 Pierre Janet; *Boston Med. and Surg. Journal*, Jan. 19, 1905, p. 93 and *Les Obsessions et la Psychasthenie*, Vol. I., p. 502.

2 Prince; *Journal of Abnormal Psychology*, 1906.

3 Babinski; *Societe de l' internat. des hopitaux de Paris*, Seance du 28 Juin, 1906.

4 Janet; *Boston Med. and Surg. Jour.*, Dec. 1906.

we may say that hysteria and epilepsy are identical, although some writers seem inclined to regard them as such. In an interesting paper Putnam and Waterman¹ discuss the relation of the hysterical to the epileptic attacks, and cite a number of cases with more or less doubtful diagnosis. They think it is possible that the paroxysms of epilepsy and hysteria, or at least, certain forms of epileptic and hysterical manifestations may stand more closely in relationship than has hitherto been assumed, and in this they do not lack the support of other writers. No one, they say, has studied the subconscious memories of epileptics, and closer resemblances may be found between these and subconscious states of the hysterics than we now imagine. They even think it may be possible that in patients with states of altered consciousness, hitherto classed as strictly epileptic, memories of the paroxysmal period might be developed through the medium of hypnosis.

There is much danger of making a mistake in diagnosis when the question is one of psychic epilepsy. Thus Wm. A. White² describes a case which resembled one of epilepsy of the psychic type. The patient had at times a condition bordering on double consciousness, and its relation to psychic epilepsy was shown by an aura preceding the attacks. White says that the real character of the case was one of mental dissociation, the presence of dissociated systems in the depths of the subconscious. In this case hypnosis and hypnoidization were employed, the details of the events for which the patient was amnesic were thoroughly traced and united to her upper personal consciousness, and a cure was effected. Probably this case would have been considered by some as one of psychic epilepsy, but it evidently was not.

A case, possibly even more interesting as regards the subject I am discussing, is reported by George M. Parker³. A man had attacks preceded by a feeling of pain and distress over the epigastrium and a foul taste in the mouth accompanied by a fetid odor. He usually fell and lay quietly outstretched, sometimes his hands fumbled aimlessly.

¹ Putnam and Waterman, *Boston Med. and Surg. Jour.*, CLIII., 509, 1905.

² *Psychopathological Researches* by Boris Sidis, Wm. A. White and George M. Parker, G. E. Stechart, New York, 1902.

At times he had general motor disturbances, his arms and legs twitching and he frothed at the mouth. Sometimes he would not fall but sat in a chair staring fixedly before him. Any degree of excitement was sufficient to initiate an attack. Complete amnesia and stupor followed all attacks. He became progressively unable to attend to his duties, constantly forgot and became confused. His physician regarded the condition as epilepsy. Parker found that the patient could now and then give a stray memory from one of the attacks, and as he says, the definiteness of the flash of recovered fragments of memory did not quite resemble the mental condition of the attacks of typical organic epilepsy. Under hypnotization he succeeded in producing a recovery of memory of the events occurring during the attacks and in tracing the origin of the foul taste in the mouth, and thereby proving that the condition was a functional psychosis simulating epilepsy. By hypnosis and hypnoidization, the latter originated by Boris Sidis, and consisting in the production of a state of abstraction, mental composure and relaxation, a cure was effected.

Parker, in speaking of this case, says it shows that many a typical epilepsy may on closer study turn out to be a functional psychosis. This is especially true of the psychic epilepsies which investigations in their laboratory have demonstrated are all pure functional psychoses, subconscious dissociated states, having the tendency to recur, periodically or not, and often mimicking the psychomotor manifestations of epilepsy, but he does not call them hysteria. The treatment in such a case, as Parker puts it, consists in bringing the dissociated memories out of the depths of the subconscious and reassociating them in the synthesis of the upper personality, restoring all the psychic material to the contracted active personal consciousness, and thus bringing about a state of former mental activity which will maintain the former synthesis. The treatment in both these cases was by hypnosis and hypnoidization.

If we are to include the psychasthenic attacks under hysteria, there is no need of making any distinction for them, and yet I think Oppenheim is right in putting them in a class by themselves. Two cases have been under my observation that have a resemblance to epilepsy, but careful

study of them has convinced me that they are neither epilepsy nor hysteria, and I report them in the hope that they may draw attention and discussion to the subject of psychasthenic attacks.

C., twenty-one years old, consulted me about five or six years ago, at which time the following notes were made :

A maternal uncle died in an insane asylum. No convulsions had occurred in the family of either parent. The mother of the patient is very irritable, easily excited, and somewhat quarrelsome. The patient has had five brothers but no sisters. His father has shown great artistic talent.

The first son is a sculptor and is irritable.

The second son does not appear to be neurotic.

The third son was very eccentric. He tried to commit suicide two or three times while at home, and finally succeeded. He at times became very much depressed and when occasionally he had these attacks he would wander away and stay away over night. Once he was absent two days. Before he wandered away his expression would become peculiar, so that his relatives would know he was about to leave home. In one attack he went a distance of several hundred miles.

The fourth son seems to be normal.

The fifth son is the patient.

The sixth son is sixteen years old and is afraid to go into the dark.

C., the patient, has never had convulsions. When he was seventeen years old he had his first visual hallucination. He was in church. He heard the minister begin his sermon, and then as he looked across the church he noticed that a certain man was looking at him. At first he liked the face and something in it reminded him of a boy of whom he had been very fond and whom he had not seen for about two years. This boy had taught him masturbation. In this first attack he did not know what occurred about him, he got up and came out of the church after the service was over, and the money he had intended to put in the plate he had still in his hand. Whether this was unconsciousness or not is uncertain. He did not speak during the attack. After this first attack he began to hate and fear the face, and always had a warning before seeing it, "a sort of spasm

would go through his whole body," or if he were holding a book his hand would tremble violently, and then if he looked across the church the "face" would be looking at him. It was always the same face and had always the same sneering expression. Except on one occasion the man was always in the same part of the church. He was not motionless, but was not seen by the patient to walk out of the church except on one occasion, when he followed the patient. During the first year the man was observed always in the same church, then he was seen in another church, and later was seen repeatedly on the street. The whole figure of the man was visible, but the patient spoke of the hallucination as "the face."

If the patient fixed his eyes upon the wall he could bring the figure of the man before him but it did not seem real to him and did not "satisfy" him, as he expressed it. The attacks occurred every Sunday during the first year, but not so frequently during the second year, and during the third year not more than four times. The patient believes the face is real. The attacks have been frequent proportionately as masturbation has been frequent.

He can always tell the day in advance that he will see the face. His eyes seem to be out of focus, he cannot keep them focused upon the model he is copying, and if he tries to do so he gets a bad headache, then becomes sleepy for the rest of the day. The aura usually occurs about 4.00 P.M. He has always had a bad taste in his mouth during the aura, "like cheese after you have eaten it the night before," or a "musty taste." If he falls asleep after the aura he has very vivid dreams, on one occasion he saw a comet coming toward him, and exploding; and on another he saw clearly the face of his brother who a few days later committed suicide. He has had the aura without seeing the face the following day, but he has never seen the face without having had the warning the day before.

Within about fifteen minutes after the visual hallucination he gets sleepy and stupid, and does not know what is going on about him. The vision is followed by fullness of the head and palpitation of the heart. He has formed the habit of taking a back seat in church so he can hide behind the people and from the vision, and support himself against the

wall. Only on one occasion he saw two children with the man. Each time he has had the vision his "eyes have gotten out of focus" and objects seem to move to and fro. He then has had a sick feeling and had to sit down. He has the same taste in his mouth during the hallucination that he has during the aura. Immediately after seeing the man his "ears ring like bells and insects all singing together." He has had occasionally aural hallucinations without other disturbances, *i.e.*, he had heard his mother calling him, or bells ringing.

The young man is very intelligent. He has never wet his clothing during an attack nor cried out, nor bitten his tongue. When he feels he is about to have the vision and resists it his face flushes, he gets cold and hot alternately, and all objects appear queer.

I have seen this patient again within the past few months. He is in excellent health. The visual hallucinations lasted one or two years after I first saw him, and then ceased entirely. They gave place to a difficulty in swallowing. The man believed he could not swallow and was depriving himself of food. An examination showed nothing abnormal to explain the dysphagia. The condition lasted about a year and a half and then disappeared.

Interesting in this report are: The neurotic history; the attacks of wandering in one brother; the aura always preceding by one day the visual hallucination and associated with a bad taste in the mouth, suggesting Hughling Jackson's uncinat group of fits, and associated also with ocular disturbances and followed by drowsiness; the occurrence of the hallucination at first always in one place, a church, and therefore in a crowd, but later in other places and on the street; the resemblance of the face seen to that of a boy who had taught him masturbation, the frequency of the attacks proportionate to the frequency of masturbation; the bad taste in the mouth and sick sensation during the attack; the possible unconsciousness in only one attack; the drowsiness, fullness of head and palpitation of heart following an attack, and the absence of all convulsions.

Oppenheim says that an abnormal impulse to wander occurs in certain individuals presenting all the symptoms of psychasthenia, who are neither hysterical nor epileptic.

One brother of my patient showed the fugue.

The occurrence of the attack for a long time only in one place, therefore in association with a certain train of thought or a certain environment, occurs sometimes in epilepsy; thus Raymond in his lectures refers to a case of psychic epilepsy in which epileptic convulsions were produced by directing the patient's thoughts in a certain channel. Worthy of note is the fact that the face seen was that of a boy with whom the patient had been friendly and from whom he had learned masturbation, and also the occurrence of the attacks in proportion to the frequency of masturbation. Freud¹ has called attention to the importance of sexual errors in the etiology of the neuroses. There is in this case something very much like a subconscious working and made evident without the use of hypnosis, and it in itself makes a diagnosis of epilepsy improbable, as does also the cessation of all attacks during a period of several years under psychotherapy. Possibly the condition may be explained, as is done for many other peculiar mental states, as a dissociation of the personality.

The diagnosis of psychic epilepsy has to be considered in this case, and indeed at first I thought it might be the correct one. The treatment was conducted some five or six years ago by a method similar to that which Dubois has recently made familiar, viz., by explaining the condition to the patient, assuring him that the attacks were not serious, and arousing his resistance to them. The result was all that could be desired, and the patient has had no attacks for three or four years. The difficulty in swallowing, evidently psychical, that followed the cessation of his attacks, is worthy of notice.

The second case has much resemblance to epilepsy and was so regarded by some, but careful examination showed that it could not be so considered.

A. B., thirty-six years of age, consulted me September 10, 1906. He was a patient of Dr. Radcliffe Cheston, and is at the head of a large business. He comes of a neurotic stock. His father had nervous prostration and now is tormented by unreasonable doubts as to the manner in

¹ Freud: *Sammlung kleiner Schriften zur Neurosenlehre*, Franz Deuticke, Leipzig und Wien, 1906.

which he conducts his business.

The patient is a hard worker. Some years ago he undertook to study a profession, but as he was engaged all day, he was obliged to study at night. After passing his examinations he "went all to pieces," as he expressed it. He would sign a letter and after a few minutes would tear the envelope open to see whether he had signed the paper. On one occasion he visited his sister, and wandered about the house in a dazed condition. He was very irritable. Two or three years ago he went away for complete rest for fourteen weeks, and since then he says he has "felt his nerves" more. He had difficulty in fixing his mind on his work, and has frequently repeated his actions in order to be sure he had done his work properly, and often would read a paper without being able to fix his attention upon it. He took exercise in the same energetic manner in which he carried on his business.

He is the father of three healthy children, two of whom are "high strung." He gives no history of sexual irregularity. He had never had any attacks of any kind before July 4, 1906.

In the early part of the summer of 1906 he went to Europe and took an automobile trip from Paris lasting two days. He traveled about two hundred miles each day. The weather was not very warm, but he was much tired by the trip. After the second day's journey, in the evening while talking to some one, he fell and was said to be unconscious. His face was a little flushed and his eyes had a vacant look. He was "perfectly limp" about ten minutes, then he became rigid in his feet, and had involuntary movements of the upper limbs of a purposive character. He was put to bed and had what he called a "chill"; he shook all over, his teeth chattered, he became red in the face, and cold in the feet, but the thermometer showed no rise of temperature.

After the first attack he had weak spells in which he would sit with a vacant expression. These attacks occurred once or twice a day and nearly every day. He was not unconscious in these attacks, although he seemed to be "wandering in mind."

The second severe attack occurred July 13, and in this

he fainted. Other attacks occurred July 17 and 21. In one of the latter he had a vacant expression, became limp, then got up and tried to walk and fell after he had gone upstairs. He then became unconscious and was rigid. After this attack was over he got up and went to bed, and had a chill and headache in bed.

Attacks occurred on July 24, 26 and 27. After the first two or three attacks he had a warning in general weakness and a sensation as if he were "charged with electricity." His memory began to be impaired. The attacks continued every few days until he got on a steamer August 29, on his return journey. He had two attacks on board the steamer. In all he had twenty-five major attacks. The last attack was on September 3. In one attack when walking alone he fell and became covered with mud.

Before taking the steamer he had an attack in which he was unconscious one hour, and in this his body was more rigid than in the other attacks. In two attacks the face twitched, but convulsive movements were confined to the face.

He had taken one drink of whiskey or beer daily, occasionally some claret, and had smoked about twenty cigarettes daily. He had also been much worried about his wife's condition as she had been in a hospital three times.

Before leaving America he had been having headaches since April, 1906, more on the right side and in the parietal region or over the mastoid process, but this pain disappeared. There was no mastoid disease.

This was the history as I obtained it from the patient and his wife at my first interview. A further study of the patient by himself revealed some interesting facts :

He told me that sometimes when he was supposed to be unconscious he was not so, and the so-called rigidity was often purely voluntary on his part, and was the result of an attempt to get the numbness out of his forearms and ankles. While he was on the ocean on his way to Europe he kept control of himself, but when he reached Paris he did not care and gave way to his feelings. He did not care whether he fainted or not, as it was a temporary relief to do so, and if he felt like sitting still and looking at a spot upon the wall he did not make a mental effort to avoid doing

it. He would become weak after dinner, and he began to dread this period, and if he could have been fooled regarding the time of day he believed he would not have fainted. He does not know why he fell.

The man presented no hysterical stigmata nor signs of organic disease. The treatment was psychotherapy.

I saw him again October 19, 1906. He had been weak twice and had had constant slight headache. On one occasion he felt that an attack was coming on, his wife urged him to resist it, but he pleaded with her to be allowed to fall, pushed her from him and said she must let him fall. His attacks have usually occurred when his wife was present. At the time of these two attacks he was again on a vacation with no business to occupy his mind, and he felt that he need not control himself. He told me that if he were asked any time in the day whether he had a headache he would say yes, and yet usually he was not aware of headache. It was not real pain that he experienced, but as he expressed it it was "consciousness that he had a head." He said if he had known that falling would have injured him he would never have fallen. Such a case as this can hardly be regarded as epilepsy, and it is questionable in my mind whether it could be regarded as hysteria.

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INTERNAL HYDROCEPHALUS.

WITH REPORT OF TWO CASES, ONE RESULTING FROM
OCCLUSION OF THE AQUEDUCT OF SYLVIVS.¹

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It is by no means rare to find occlusion of the aqueduct of Sylvius as one of the etiological factors in internal hydrocephalus mentioned in text-books dealing with this subject. Attention is particularly attracted by the fact, however, that no references are given, in most of the articles which refer to this occlusion as a possible cause, whereby one can determine that the author is speaking from clinical experience and not theorizing.

We have been struck with the *a posteriori* nature of discussions on this subject, and it is therefore with pleasure that we bring forward a case of internal hydrocephalus the cause

¹ This article also appeared in the Journal of the American Medical Association, April 13, 1907.

of which is probably congenital, almost total occlusion of the aqueduct of Sylvius.

Anton,¹ in dealing with the causes of hydrocephalus, quotes Baginsky's opinion that acute hydrocephalus is identical with acute non-tuberculous inflammation of the choroid plexus and the following increase of exudation. He adds that there is without doubt involvement of the ventricle walls and the ependyma. He also advances the view held by Rindfleisch, Huguenin, and von Bönninghaus to the effect that the inflammatory affection of the ventricles and choroid plexus is always associated with a meningitis serosa or purulenta, and that these are caused by toxic or infectious factors. Referring to Macpherson, he states as a possible cause an infectious phlebitis with thrombus formation of the venous sinuses. He says that occasionally the exciting agent of the inflammation gains access directly to the ventricle through the brain substance, with or without traumatism, and there sets up a fibrinopurulent process. Lastly, he calls attention to a possible connection between rachitis and hydrocephalus.

Gowers² holds that the "only known cause" of acute hydrocephalus is meningitis. Speaking of chronic internal hydrocephalus he makes this interesting statement: "Sometimes all the ventricles are distended; more often the fourth ventricle suffers little, even where there is no obstruction in the aqueduct of Sylvius. In such a case the aqueduct may be funnel-shaped, dilated toward the enlarged third ventricle." This statement should be compared with the gross anatomical findings of our second case. Gowers further mentions the possible obstruction of the veins of Galen preventing the return of venous blood from the intraventricular vessels and thereby becoming a cause. Such a condition as this, if acute, is likely to cause ventricular hemorrhage and rapid death, as in a case reported by one of us (Spiller) with Wadsworth,

¹ Flatau-Handb. path. Anat. d. Nervensystems.

² Dis. of the Nervous System.

but when developing slowly might be productive of internal hydrocephalus.

Oppenheim¹ speaks of compression of the aqueduct of Sylvius as a cause of internal hydrocephalus. D'Astros² states that an obstruction of the aqueduct of Sylvius will produce *une hydropsie* of the third and lateral ventricles; but he gives no instance of its so doing. A Henle³ says that a closure of the aqueduct of Sylvius will cause a hydrocephalus confined to the third and lateral ventricles; but he gives no reference to a case. P. Bourneville⁴ speaks of "another case" of hydrocephalus caused by the obliteration of the aqueduct, but no further information is given. F. Schultze⁵ mentions the possibility of hydrocephalus being caused by the obstruction of the aqueduct, and refers to Seeligmüller's cysticercus case which we cite below.

Seeligmüller⁶ speaks of a possible pressure on the aqueduct of Sylvius by a tumor causing hydrocephalus. In the same work he gives the history of his case of hydrocephalus caused by the plugging of the aqueduct by a cysticercus the size of a cherry stone. Apropos of cysticercus Karl Osterwald⁷ has collected about 34 cases in literature of cysticercus in the ventricles of the brain, but in none of the cases given was the parasite in the aqueduct.

Bönninghaus⁸ gives as a cause of hydrocephalus, what he terms an "acute automatic closure" of the aqueduct of Sylvius. By this he means that when the effusion into the third and lateral ventricles occurs rapidly, the more yielding portions of the third ventricle, *i.e.*, the floor and the roof, become dis-

¹ Lehrb. d. Nervenkrankheiten.

² Les Hydrocéphalics, 1898, 75

³ Beitr. z. Path. u. Therap. d. Hydrocephalus.

⁴ Brouardel, Traité de médecine.

⁵ Nothnagel, Specielle Pathol. u. Therap.

⁶ Krankheiten d. Rückenmarks u. Gehirns.

⁷ Beitr. z. d. Diag. Cysticercus Ventriculi Quarti, Neurolog. Centrbl., March 16, 1906.

⁸ Die Meningitis Serosa Acuta, 1897.

placed and pull from above and below on the more solid pons, and in this way produce an elongation of the aqueduct which may result in complete closure.

One of us (Spiller¹) reported a case in which the third and lateral ventricles were dilated and the aqueduct was occluded by proliferation of the neuroglia. Two other cases of partial internal hydrocephalus have come under his notice. In one the distention was confined to one lateral ventricle of the brain and was caused by partial inflammatory closure of the foramen of Monro; in the other case only the posterior horn

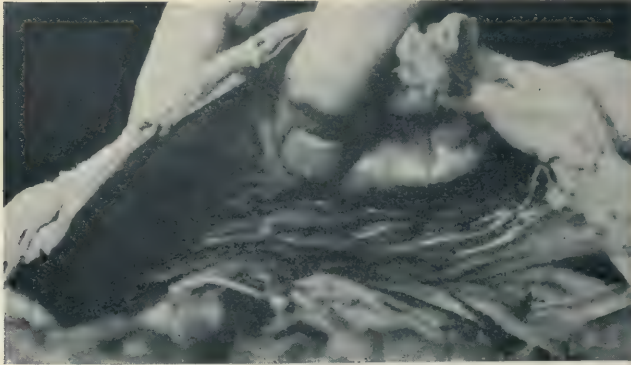


FIG. 1.—Photograph of one cerebral hemisphere, showing the great distention of the lateral ventricle and the thinness of the brain tissue.

of the lateral ventricle was distended, and very greatly, from unknown cause. In this case the occipital lobe of the affected side was a mere sac. He also refers to a case of Bourneville and Noir² in which the aqueduct was entirely occluded, but there was no microscopic study of the tissues. In this same paper he (Spiller) speaks of Touche's case,³ in which there was occlusion of the aqueduct.

Our attention has been called by Dr. H. E. Becker, of Quincy, Ill., to a case of occlusion of the aqueduct reported

¹ Amer. Jour. of Med. Sci., July, 1902.

² Le progrès méd., July 14, 1900, 17.

³ Bull. et mémoires de la Soc. méd. d. hôp. de Paris, 1902, vii, 141.

by Hilton in his *Lectures on Rest and Pain*. The sections from still another case (unreported) have been shown us by H. S. Hutchinson.

Our first case was reported by one of us (Spiller) without microscopic study.¹ The history as obtained from the Pennsylvania Training School for Feeble-minded Children is as follows:



FIG. 2.—Photograph of a child, aged one and a half years, with intense hydrocephalus.

Patient. Sally H., aged sixty-two years at time of death; American.

History. Father died of consumption, aged thirty-nine years; mother died aged sixty-three years, cause unknown; two brothers and two sisters died of consumption. One sister (the first born) died in early infancy, cause unknown; two brothers are living in good physical and mental condi-

¹ Jour. of Nervous and Mental Dis., 1900, 559.

tion. Of the details of the patient's birth nothing is known. She was supposed to have been born hydrocephalic and was weak and sickly as a baby. Sight, hearing, and speech were good. She had epilepsy for years and during her seizures would froth at the mouth and purge. She suffered frequently from vomiting without assignable cause, and had occasional attacks of vertigo. She was said to have chronic kidney disease six years prior to her death. There was no paralysis at any time, and although no joint complication was present she always walked as though lame. She was said by Dr. Llewellyn, who had known her for years, to have been fairly well developed mentally. Her advanced age and her mental condition are remarkable in view of the extensive hydrocephalus.

Macroscopic Examination. The third and lateral ventricles of the brain were greatly dilated and the white matter surrounding them was intensely atrophied. The whole cerebrum appeared as a greatly distended sac. The lateral ventricles freely communicated with one another, and the walls of the third ventricle were so pushed apart that the floor of this ventricle was on a level with the upper part of the basal ganglia.

Microscopic Examination. Left optic thalamus: The blood-vessels were thickened and there were some recent hemorrhages within the substance of the thalamus. All the vessels were distinctly congested. There was little vacuolation of the cells of the choroid plexus such as described by Raubitschek.¹

Corpus callosum: A section from the median portion of this structure was not more than 1 mm. in thickness; it stained normally and contained numerous amyloid bodies.

Left occipital lobe: In places the cortex and white matter were not more than 2 mm. in thickness; in other places the thickness was 6 mm. The atrophy was relatively much

¹ Ztschr. f. Heilkun., 1905, p. 219.

greater in the white substance than in the cortex. Where the width of the section was the least, more than half was occupied by the cortex. The cells were much diminished in number, as shown by the thionin method.

The fornix: This was much atrophied and the bloodvessels within it were greatly thickened.

The aqueduct was almost occluded, this occlusion being probably a congenital malformation because the cavity was well lined throughout by a layer of ependymal cells, which would hardly be the case if the occlusion were caused by proliferation of the neuroglia. The neuroglia tissue immediately adjoining the ependymal lining was a little proliferated, and the aqueduct was filled with red blood corpuscles which were unaltered in shape, and probably were deposited there shortly before death. To the naked eye the aqueduct seemed to be entirely occluded. In a microscopic section the aqueduct to the naked eye was hardly as large as the point of an ordinary pin.

The second case is reported as one of intense congenital hydrocephalus without occlusion of the aqueduct. The pathological material was received from Dr. Thompson S. Westcott, to whom the case belongs. The patient lived to be a year and a half old. The skull was excessively large. It is difficult to describe the gross appearance of the brain other than to say that it was a large, thin bag of nerve tissue so attenuated in many places as to be translucent, and the mere handling of it caused numerous tears in its substance.

Microscopic Examination. The corpus callosum was not more than 0.5 mm. thick.

Left occipital lobe: Sections in places were not more than 1 mm. thick. Here, as in the other case, the atrophy was much more pronounced in the white matter than in the cortex.

Left motor area, median side: The white matter was most affected. There was great diminution in the number of cells and the pericellular spaces were all distended. The cells appeared all decidedly smaller than normal. The pia was

not thickened and the cortex and white matter were 4 mm. thick.

In contrast to the other case the fourth ventricle was greatly dilated as the result of the dilatation of the aqueduct of Sylvius. The cause of the hydrocephalus in this case could not be determined, but the condition seemed to have been a congenital malformation.

The two cases show very clearly that the distention of the fourth ventricle may depend on the patulency of the aqueduct of Sylvius.

CONJUGATE DEVIATION OF THE EYES AND HEAD AND DISORDERS OF THE ASSOCI- ATED OCULAR MOVEMENTS.

IN TUMORS AND OTHER LESIONS OF THE CEREBRUM.*

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VARIOUS VIEWS DISCUSSED.

Within the past two years the causes of conjugate deviation of the eyes and head have repeatedly been the subject of controversy, especially by certain French writers. In 1904, Bard¹ advanced the sensorial theory of conjugate deviation, although this had been spoken of previously by Roux.² This theory has since been ably supported, especially by Dufour,³ and opposed by Grasset,⁴ Dejerine and Roussy,⁵ and others.

Chiefly through the kindness of Dr. Charles K. Mills and Dr. William G. Spiller I have had the opportunity to study 16 cases of conjugate deviation of the eyes and head, 15 of which are with necropsy, and I have attempted to come to such conclusions as to the causes of conjugate deviation of the eyes and head as the evidence in these cases will permit, together with the aid of cases in the literature.

Disorders of associated ocular movements occurring in lesions of the cerebrum will be discussed only in so far

* Read in the Section on Nervous and Mental Diseases of the American Medical Association, at the Fifty-seventh Annual Session, June, 1906.

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1. Bard: *Semaine Médicale*, Jan. 13, 1904.

2. Roux: *Arch. de Neurol.*, September, 1899.

3. Dufour: *Rev. Neurol.*, April 15, 1904.

4. Grasset: *Rev. Neurol.*, July 15, 1904.

5. Dejerine and Roussy: *Rev. Neurol.* No. 3, Sept. 15, 1905.

as they occur in association with conjugate deviation of the eyes, and in cases of hemianopsia due to peripheral or central lesions. The importance in clinical diagnosis of paralysis of associated movements of the eyeballs has been discussed only recently by Spiller.⁶

The literature is replete with instances of conjugate deviation of the head and eyes as a result of lesions in almost every part of the brain. Experimental investigations on lower animals confirm what has been said of the pathologic evidence, for it has been shown that electrical excitation of any portion of the brain, if of sufficient intensity, is productive of deviation of the eyes, or of the head, or both.

There is good reason for this. Perhaps no function is more highly specialized than that which is concerned with the movements of the head and of the eyes. These are constantly moved for one reason or another; the auditory and the visual senses probably playing the most important part in the determination of these movements. Such specialization of function necessarily needs and acquires a most complex cortical representation.

With our present knowledge of cortical representation we should expect a motor center for the movement of the head, a separate center for the eyes, and still another for the combined movements of the head and eyes. Within the past few years the cortical centers for all, or nearly all, motor function have been placed in front of the central fissure. This has been borne out by experimental and pathologic evidence. We should expect, therefore, that there should be but one motor center for each function, and that center anterior to the Rolandic fissure. It has been shown that movements which are performed conjointly by corresponding or correlated muscles on both sides of the body have a cortical representation in each cerebral hemisphere. Therefore we should expect a bilateral cortical representation for the deviation of the eyes and of the head.

Any auditory, visual, or olfactory stimulus will cause conjugate deviation. Touch, pain, or temperature stimuli will also cause deviation. Besides it has been held that reflex or automatic and voluntary impulses initiated independently of these senses may cause such deviation.

6. Spiller: *Jour. Nervous and Mental Dis.*, July and August, 1905.

Taking up the subject of conjugate deviation of the eyes or of the head, or of both, as the result of anyone of the above-mentioned sensory stimuli, what is the cerebral mechanism of this conjugate deviation? Illustrating by the sensation of hearing, we presume that an auditory impulse is first transmitted to the auditory center in the temporal lobe, from here it is carried by means of association fibers to the motor center, from which motor impulses are transmitted to the nerve cells which supply the muscles concerned with the function to be performed. In like manner a unilateral motor center for the conjugate deviation of the eyes and head should be in intimate connection by means of association fibers, not only with the auditory center in the temporal lobe, but also with the visual center in the occipital lobe and with the olfactory centers in the uncinate gyrus and the gustatory center wherever situated, and with the centers for touch, pain and temperature senses in the parietal lobe. In brief, sensory irritation in any portion of the body will cause deviation of the head and eyes in the direction of the irritation through the correlated activities of the sensory and motor centers concerned. Not only that, but the centripital fibers which transmit these impulses from the periphery to the cortex must be intact for the proper interpretation of these stimuli; and the centrifugal motor tracts must also be intact for the physiologic performance of their functions. Should any portion of this most complex mechanism be disturbed, the result would be failure in normal deviation of the head and eyes, the deviation depending entirely on the nature and location of the lesion.

The most recent writers on this subject, Cantonnet and Taguet,⁷ advance the idea of an automatic or reflex center for ocular movements in the optic thalamus. In support of this they quote cases of cortical lesions in which there was paralysis of voluntary ocular movement, but in which reflex or automatic movements were preserved. We have always regarded reflex movements of the eyes as best illustrated by the fact that the eyes will follow the moving finger or hand in different directions; and that similar deviation will take place in response to various sensory stimuli. It would, perhaps, be advisable if we were to define "reflex" and "auto-

7. Cantonnet and Taguet: *Rev. Neurol.*, No. 7, 1906, p. 308.

matic" movements more clearly. When the eyes deviate in a certain direction because of a noise, that is a reflex action, only in the sense that a movement is performed because of a sensory stimulus. If the sensory part of the reflex arc is disturbed, as in deafness, no amount of noise will cause deviation. Why should this be called an automatic movement?

It has been shown that in lesions of the thalamus, we sometimes have such symptoms as forced or involuntary laughing or crying, and in lesions of the superior cerebellar peduncle there may be incoördinate movements of some of the limbs. These are examples of automatic movements. If we assume that a similar automatic center exists for ocular movements in the thalamus, should not a disturbance of such a center cause forced or incoördinate ocular movements?

In his very able presentation of the subject of associated ocular movement, Spiller¹⁰ agrees with others that paralysis of associated ocular movement laterally is due to lesions of the posterior longitudinal bundle, and asserts that a paralysis of upward and downward movement is probably the result of a lesion near the oculomotor nuclei. He believes that a tract similar to the dorsal longitudinal tract, and possibly a part of this fasciculus, situated near the oculomotor nuclei, connects these nuclei, and that an interruption of the tract causes failure of associated upward or downward ocular movements. He does not admit the need of a basal coördinating center for ocular movements, near the oculomotor nucleus.

When the eyes follow the finger, this is really an associated movement, and the result, not of a pure reflex action, but of a distinct sensory (visual) impression, in the same sense that a noise (auditory impression) will cause deviation of the eyes. If the sensory arc is interfered with as in blindness, the eyes will, of course, not be able to follow the finger as the result of a visual impression.

A voluntary movement is the result of a stimulation of a motor center. The question arises whether in a normal individual a pure voluntary impulse is possible without a previous sensory stimulation, the term sensory stimulation being used here in its broadest sense. When we will to move the eyes independently of any immediate sensory stimulus, as when in meditation, there is at least

a psychic stimulus to the cortical oculomotor center (that is, the center for the movements of the eyeballs), this stimulus may come, for example, from the higher psychic areas of the prefrontal lobe, but it is intimately dependent on stored impressions at one time or another received or accumulated by means of sensory mechanism, and moreover in the act of using the eyes in such circumstances as when one wills to read a book or to look at the stars, sensory impression takes part in the lateral and upward movements. It is probable, therefore, that a centrally initiated voluntary impulse is dependent on the functioning of a central reflex arc, and that any interference with this would cause deviation.

Summing up the views so far advanced, it is my opinion that the movements of the head and eyes are dependent on a most complex mechanism; that there is only one true or at least functionally important cortical motor center anterior to the central fissure; that this motor center is in immediate connection with the sensory portions of the brain; that whenever the head and eyes are moved it is the result of a sensory stimulation, and that any interference with the sensory-motor arc will cause impairment of this function. Let us now consider in order the embryologic, anatomic, physiologic and pathologic evidence in support of these views.

EVIDENCE IN SUPPORT OF VIEWS.

The well-known myelogenetic investigations of Flechsig⁸ support these views. According to him, in his primordial zones or regions of early development are included exclusively projection fibers, sensory and motor, or, as he terms them, corticopetal and corticofugal. He believes that for every sensory path there is a corresponding motor path. In the primordial zones are included the motor and sensory regions, the visual or occipital lobes, the auditory or upper temporal regions and the olfactory and gustatory centers principally in the anterior temporal region. The myelogenetic differences between the primordial and the intermediate and terminal zones, according to Flechsig, are the result of successive development of different sense organs and their motor or centrifugal paths, and the development of long and short association systems.

8. Flechsig: "Gehirn und Seele." Leipzig, 1896.

In the main these views have received physiologic and pathologic confirmation. What appears to be the view of Flechsig that each primordial area is both sensory and motor can not be altogether agreed with, but it must be recalled that his views are only relative, for in his myelogenetic area No. 1, which corresponds with some closeness to the central convolutions, the tactile radiation is distributed mostly to the postcentral convolution, while the tract which is undoubtedly motor originates from the precentral convolution. This would argue for separate cortical representation for motor and sensory functions, and would accord with our present views.

With the auditory or cochlear myelogenetic area in the first temporal convolution Flechsig finds a corticofugal radiation, which he believes may be motor, although this has not as yet been demonstrated. The cells and fibers which constitute this radiation lie at the lower border of the first temporal and in the furrow which separates it from the second temporal convolution. He assumes that motion of the head and of the body following an auditory impression may be performed through the intermediation of corticofugal fibers which originate in or near this center. Similarly, Flechsig reasons that movements of the eyes and of the head consequent to visual stimulation are due to the intermediation of corticofugal fibers from or near the visual zone. Motion as a consequence to olfactory stimulation can likewise be explained.

According, then, to Flechsig's view, a movement of the eyes or of the head is the result of a sensory stimulation and may be due to an auditory, visual or other sensorial impression coming by way of the corticofugal fibers which originate in these centers. I do not agree, however, with the view that the corticofugal fibers coming from the special sense centers are purely motor projection fibers, as Flechsig appears to believe. Undoubtedly the corticofugal radiations associated with the corticopetal radiations in Flechsig's myelogenetic area No. 1, which corresponds to the sensory and the motor centers around the central fissure, are purely motor. The corticofugal radiations of the primary auditory and visual zones are probably not purely motor projection fibers, but in large part are fibers which connect these special sense areas with the oculomotor centers in the

main motor region. A sudden auditory stimulation will, of course, cause immediate turning of the head and eyes, but this is not because, or at least not usually because, a motor impression is transmitted by the corticofugal fibers directly from the auditory center to the nuclei which are concerned with the muscles necessary to perform that function, but rather because the auditory impression is transmitted by means of association fibers to the motor cortex and thence to the basal nuclei.

When an auditory impulse is received in the temporal convolution it may be stored up in the auditory memory center, it may be transmitted to the visual center and recall visual images associated with the sound produced, it may cause olfactory or gustatory stimulation or it may cause the movement of almost any portion of the body as the person wills. In the performance of all these possible functions it is necessary to have the auditory center in direct communication with the other sensory centers, and this is accomplished by means of associated fibers, the existence of which have been repeatedly demonstrated; similarly the visual centers are likewise in association with other centers.

Mills⁹ believes that each sensory area has its motor correlative and that each of these has its separate cortical representation. He speaks of a sensory motor, visual motor, auditory motor, gustatory motor, olfactory motor and a possible equilibratory motor area. According to Mills, this conception does not do away with the importance of our old-time motor area, which still remains the great or main motor region. His view is that at least in the highly developed human being the centers for movements of the head and eyes and for other movements have largely taken the place of the motor projection fibers which exist in, or in close contiguity with, each of the special sense areas; in other words, the visual motor area, auditory motor area, olfactory motor area and gustatory motor area are largely, although probably not altogether, in abeyance in movement, although still retaining their importance to a greater or less degree in the animal scale below man. If this be the case, the arguments brought forward in this paper which are based on the idea of a single active functioning region for movements of the head and eyes are valid; in other

9. Mills: "Brain," 1889, and Univ. of Penna. Med. Bulletin, May, 1904

words, we can disregard all cortical motor areas except those which lie in front of the central fissure.

Experimental investigations also confirm my views, for it has been shown that electrical excitation of any portion of the brain, if of sufficient intensity, is productive of movements of the head and eyes. Four principal areas have been found in the brain of lower animals in which electrical stimulation will produce these movements: in the frontal lobe, in the so-called neck or head region, in the angular gyrus and in the occipital lobe. Partly because of the fact that stimulation of the posterior portion of the second and third frontal convolutions and the region near the angular gyrus is always productive of movements of the head and eyes, these two areas have been considered by various authors as the motor centers for these functions.

Landouzy¹⁰ and Wernicke¹¹ were among the first to favor the area near the angular gyrus or the *pli courbi* of the French. The physiologic experiments of Luciani and Tamburini,¹² Ferrier,¹³ Munk¹⁴ and Obregia¹⁵ supported their views, for stimulation of this region constantly caused deviation of the eyes and head. Against this view, however, were the extirpation experiments of Brown and Schäfer¹⁶ on the brains of apes in which conjugate deviation was not obtained. Again Flechsig has shown that this area contains few projection fibers. It is probable that electrical stimulation of the occipital and parietal regions causes deviation of the eyes and head, not because of a motor center in these areas, but because of stimulation of the visual and auditory fibers, respectively. It has been shown that underneath the angular gyrus lie the fibers from the occipital and auditory lobes, and it is because of the combined involvement of these that any stimulation or lesion of this area causes such constant deviation of the head and eyes.

10. Landouzy: Thèse, de Paris, 1876.

11. Wernicke: Arch. f. Psychiatrie, 1888, vol. xx, p. 243.

12. Luciani and Tamburini: See Klaas' Dissertation, Marburg, 1898.

13. Ferrier: "Hirnlocalisation," 1890, Leipzig and Vienna.

14. Munk: "Sitzungsberichte der Kgl. akad. der Wissenschaften zu Berlin," 1890.

15. Obregia: Archiv. f. Anat. u. Physiol., 1890, p. 260.

16. Brown and Schäfer: Phil. Trans., Royal Society of London, 1888.

The work of Ferrier,¹³ Mott and Schäfer,¹⁷ Beevor and Horsley,¹⁸ and lately of Sherrington,¹⁹ has conclusively proved the existence of an oculomotor center in the posterior portion of the second and third frontal convolutions. The existence of a separate center for the movement of the head, for the eyes and a combined center for the movement of the head and eyes has been repeatedly shown in lower animals, and it is probable that such differentiation also exists in man. Beevor and Horsley¹⁸ in apes (*Macacus sinicus*) found an isolated center for movement of the head, for the eyes and for the head and eyes in the posterior portion of the first, third and second frontal convolutions, respectively. Later these authors found in an orang-outang, in the precentral convolution, underneath the hand region, a center in which irritation caused deviation of the head and eyes, and, on the other side of the brain, deviation of the eyes alone because of irritation of the foot of the second frontal convolution.

A critical study of the evidence furnished by the physiologic experiments in lower animals favors the view that there is a separate center for the movement of the eyes in the foot of the second frontal convolution, that a separate center for the movement of the head exists in the lower part of the precentral convolution along its anterior edge, and that further stimulation of either area may cause a combined deviation of the head and eyes.

Normally, the head and eyes are moved together more often than separately, and the eyes more than the head in separate movement. We should expect, therefore, from developmental and physiologic reasons, that the oculomotor center should be better developed than the head center, and that stimulation of the head center should more often give conjugate movement of the head and eyes than stimulation of the eye center. This is proved by the experiments of Beevor and Horsley,¹⁸ Ferrier¹³ and Mott.¹⁷ The experiments of Sterling²⁰ are of extreme interest in this connection. This author irritated the so-called neck region in young animals and found that he could obtain movements of the head alone

17. Mott and Schäfer: "Brain," 1890.

18. Beevor and Horsley: Phil. Trans., Royal Society of London, vol. clxxxi, 1890, p. 129.

19. Sherrington: Jour. Phys., 1894.

20. Sterling: Arch. f. Anat. u. Physiol., Phys. Ab., 1903, p. 486.

in dogs 8 days old, whereas associated movements with the eyes could not be obtained until the twenty-first day. He did not perform the converse experiment by irritating the ocular centers alone, which would be interesting. We can, however, argue from this that separate centers for the eyes and head exist, and that because of association between them irritation of one may cause movement of both. It is also for this reason that irritation of other portions of the brain besides the motor, cause in most instances combined movement of the eyes and head.

Our knowledge of the direction of the movements of the eyes and of the head is chiefly in regard to lateral deviation. Reasoning from analogy, we should expect a separate center for upward and downward movement, as well as for lateral movement. The experiments of Mott¹⁷ proved that this exists in lower animals. By irritating, in apes, the posterior portion of the frontal convolution he obtained movement of the eyes in a horizontal direction, to the opposite side and upwards, and to the opposite side and downwards, the points irritated lying from the median to the lateral side, laterally, on the outer side and in the median portion, respectively. A similar order was present for the movement of the head. Schäfer was enabled to obtain somewhat similar results by stimulating different parts of the occipital lobe.

Permanent paralysis of upward or downward associated ocular movement is only rarely observed clinically, and, as Spiller has shown, is probably due to lesions near the oculomotor nucleus. In pathologic conjugate deviation of the eyes the deviation is sometimes laterally and upward or downward, as in Case 7 of my series; but I know of no reported case in which the conjugate deviation of the eyes was purely either in an upward or downward direction. There are several physiologic reasons for this. In the first place, lateral movements of the eyes are better developed than upward movement, but perhaps no better than downward movement. There is still another important factor: One side of the brain controls movements of the eyes and head in the opposing direction; the tendency for deviation would, therefore, naturally be in a lateral direction. This was well shown in the experiment of Mills,⁹ who faradized in man the head center and obtained deviation of the head to the opposite side.

The functions of convergence and divergence are considered by the majority of authors as reflex phenomena with possibly basal centers. This I believe, however, to be an error. I am of the opinion that they are as much muscular movements and dependent on a similar mechanism as either lateral, upward or downward associated ocular movement. In convergence the internal recti muscles of either side are used, while in divergence the external recti are called into action. Should, for instance, an internal rectus be paralyzed, the corresponding eye will not be able to converge. This can be compared to failure of associated lateral movement of one eye, if the external rectus is paralyzed.

If we believe, as no one doubts, that there is a connection between the external rectus muscle of one side with the internal rectus of the other by means of the posterior longitudinal bundle, and if, as Spiller believes, there is a similar tract connecting the nuclei of the superior rectus and inferior oblique muscles of one side with the corresponding nuclei of the other side, and if this bundle is concerned with upward movement, and if there is another tract for downward associated ocular movement, why should not there be a similar tract connecting the internal recti nuclei and another connecting the nuclei of the external recti, these having to do with convergence and divergence?

It is true that the existence of such tracts has not been demonstrated, but neither have the tracts which are supposed to be concerned with upward and downward movement. There are, however, clinical facts which support this view. In the majority of cases of paralysis of associated ocular movement due to pontile lesions, either convergence or divergence is lost. In the statistics collected by Spiller, of 47 cases, the condition of convergence was noted in 26. Of these, convergence was impaired in 15 cases, and in 9, normal or nearly normal. In most of the cases in which convergence was said to be normal, the observations were made some time before death, and this function might well have been lost later.

It is more than probable that convergence and divergence have also cortical representation and that this is in the posterior portion of the second and third frontal convolutions. This subject will be fully discussed in a subsequent paper on paralysis of associated ocular movements due to basal lesions.

Let us now consider the pathologic evidence. Con-

jugate deviation of the head and eyes may occur as a result of lesions causing hemiplegia, in tumors of various localities, in areas of softening, in meningitis, in epilepsy; in fact, it can be shown that any lesion in any portion of the brain may cause deviation. Consider, for instance, the statistics collected by Grasset.²¹ Of 104 cases, 48 with necropsy, in 2, lesions were shown in the frontal lobe; in 10, in the frontal and other areas; in 25, in the internal capsule and cerebral peduncles, and in 11 the lesions were diffuse.

In the 16 cases with necropsy reported in this paper, lesions were found in almost every portion of the brain. In Case 1 an area of softening was found in the frontal lobe. In Cases 3 and 4 there were diffuse cortical hemorrhages in the frontal, parietal and occipital lobes. In Case 5 a hemorrhage was found directly in the angular gyrus. In Case 6 a hemorrhage was found in the occipital lobe and an area of softening in the precentral convolution. In Case 7 (Fig. 1) a large fibrosarcoma occupied the whole of the occipital lobe. In Case 8 there was a thrombosis of the middle cerebral artery. In Cases 9, 10 and 11 there were hemorrhages in the posterior limb of the internal capsule and the neighboring parts. In Cases 12, 13 and 14 there were lesions in the lenticular nucleus and external capsule. In Case 15 there was a lesion of the lenticular nucleus alone. In Case 16 (Fig. 2) there was an area of softening involving the anterior limb of the internal capsule and lenticular nucleus, with a fresh hemorrhage underneath the angular gyrus. Besides in Cases 14 and 15, in which no fresh hemorrhages were found, there was dilatation of the lateral ventricles.

If we consider more minutely the location of the lesions causing conjugate deviation, we find three principal areas involved: 1. The frontal area. 2. The area of the angular gyrus. 3. The internal capsule. We can still further subdivide these regions. In the frontal areas there are recorded cases of lesions involving the posterior part of the second (Sahli,²² Drummond²³ and Oppenheim²⁴) and others of the posterior part of the second and third frontal convolutions (Klaas,²⁵ Case 4 of my series, and others). Besides, there are a large number of recorded cases of lesions involving the pos-

21. Grasset: "Des Localisations dans les Maladies Cérébrales." Montpellier, Paris, 1880, pp. 215 to 238.

22. Sahli: Deutsch. Arch. f. klin. Med., vol. lxxxvi, No. 1-3, p. 1.

23. Drummond: Lancet, 1887.

terior portion of the second and third frontal convolutions, the adjacent precentral convolutions and other areas, as in the cases collected by Müller²⁶ and Klaas.²⁵ The important factor is that in all of these cases the posterior portion of either the second or third frontal convolutions or the adjacent part of the precentral convolution was involved.

Of the lesions in the region of the angular gyrus may be included any lesion in or near this area. Whatever in this region causes conjugate deviation must cause interruption of the fibers underneath the angular gyrus.

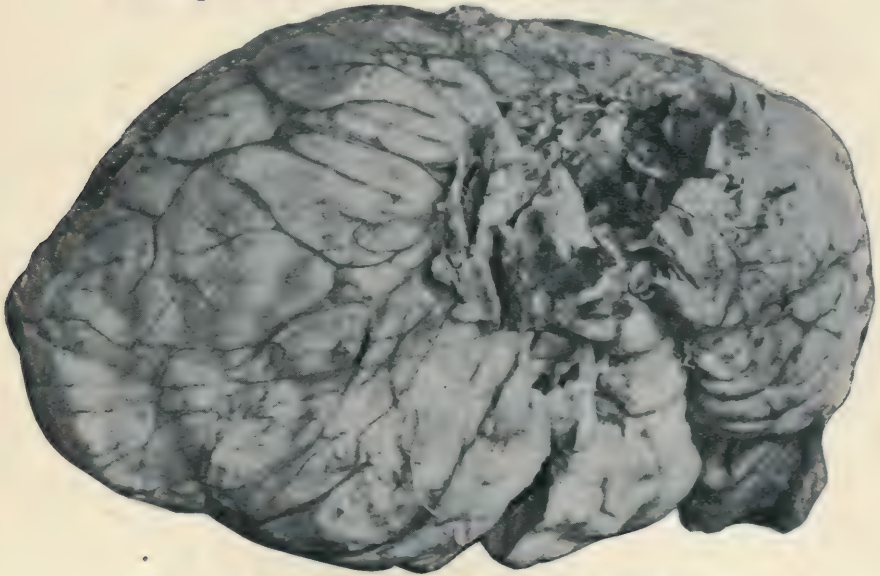


Fig. 1.—A fibrosarcoma of one occipital lobe, causing besides other symptoms, paralysis of associated ocular movements, probably as the result of the hemianopsia (Case 7).

If the lesion is posterior to this point, as in Case 6, there is only an interruption of the fibers from the occipital lobe. If the lesion is directly in the angular gyrus or beneath it, as in Cases 5, 12, 13 and 14, there is an interruption of the fibers both from the occipital and temporal lobes. In every one of these cases the

24. Oppenheim: "Charité Annalen," 1885.

25. Klaas: Inaug. Diss., Marburg, 1898.

26. Müller: Deutsch. Zeitschr. f. Nervenhk., 1902, vol. **xii**, p. 375.

lesions extended underneath the cortex and must have involved the fibers coming from both these lobes.

In the lesions involving the internal capsule, a differentiation can be made between those involving the anterior limb and those involving the posterior limb. In the former the projection fibers from the frontal lobes are involved, while in the latter there may be an involvement of the motor, sensory or optic fibers. It is the opinion of some French writers, notably Bard,¹ that to cause deviation there must always be an involvement of the optic radiations. This, however, is not correct, as in most of the recorded cases, also in mine, there was no such involvement.

From the pathologic evidence, therefore, the conclusion must be drawn that lesions involving the posterior portions of the second and third frontal convolutions and the adjacent precentral convolution cause conjugate deviation of the eyes and head, and that when lesions elsewhere in the brain produce conjugate deviation there is an interruption either of the projection fibers in or near the internal capsule or of the association fibers between the occipital and temporal cortex and the frontal lobe.

Consider now the pathologic evidence regarding the oculomotor center in the frontal lobe. It is the opinion of some, as of Sahli,²² that in man there is no evidence that a separate center for the movement of the head and eyes exists. Müller,²⁶ on the contrary, believes that there are separate centers for these functions, and he places both in the posterior portion of the second frontal convolution, the center for movement of the head being in the under portion and that for the eyes in the upper portion of this convolution. From the pathologic evidence at hand, no one doubts the existence of the oculomotor center in the posterior portion of the second and probably the third frontal convolution. The principal reason why lesions of this area cause deviation also of the head is because the head center is so intimately associated with the oculomotor center that an irritation or lesion of one will most likely cause involvement of the other.

That a separate center exists for the movement of the head was beautifully shown in the faradization experiment of Mills⁹ in man, in which a faradic irritation in the lower precentral convolution caused movement of the

head alone to the opposite side. This is proven also in Case 6, in which an area of cortical softening was found in the lower precentral convolution, causing Jacksonian convulsions limited to the face and partly to the hand, and also causing deviation of the head to the other side, but the eyes were not moved. Therefore, the center for movement of the head in the lower part of the precentral convolution should be placed below and probably a little forward of the hand center.

It is more than probable that the cortical oculomotor center in the posterior portion of the second and third frontal convolutions is subdivided for lateral and upward and downward movements. According to Parinaud,²⁷ the center for upward movement is in the lowest portion of this area; for downward movement in the upper, and for lateral movement in the median portion.

Mills and Frazier²⁸ obtained forward movement of the head by irritating the anterior portion of the head center in the precentral convolution. In another case in which the records have been lost, Dr. Mills distinctly remembers obtaining upward movement of the eyes alone. This most important evidence proves that cortical centers for upward and downward movements for the head and for the eyes exist. It is also probable that a separate center exists for the combined movements of the head and eyes, and this center is probably between the individual head and eye centers. This center may also be subdivided for lateral and upward and downward movements.

DIAGNOSTIC VALUE OF CONJUGATE DEVIATION.

So far we have not considered conjugate deviation in a clinical sense, or its value in diagnosis. Conjugate deviation of the eyes and head occurs most often in hemiplegia, and its occurrence is considered an unfavorable sign, as most such cases end fatally. Also it is most always accompanied by unconsciousness. Indeed, Grasset believes that it always is. The conjugate deviation involves mostly the head and eyes together, but it rarely involves the eyes alone, as in Cases 4, 12, 13, 14, 15, and in exceptional cases the head alone, as in

27. Parinaud: *Arch. de Neurol.*, vol. v, No. 14, 1883.

28. Mills and Frazier: *Univ. of Penna. Med. Bulletin*, July, August, 1905.

Case 6. More rarely still, the deviation of the head is in one direction and the eyes in the other, as in Case 1. The only similar cases recorded are by Roussy and Gauckler,²⁹ Dufour (cited by Roussy and Gauckler²⁹), one case of Prevost's (cited by Grasset²¹) and a case of Gaussel.³⁰

The deviation is never permanent and, as a rule, does not persist longer than two or three weeks and in most cases about a week. The deviation of the head is not so persistent as that of the eyes and is the first to disappear. At times, instead of there being a complete deviation there are jerking movements of the eyes, these being mostly in a lateral direction, but occasionally as in Case 13 they are in a lateral and upward direction. The jerking movements of the eyeballs are always from the median line to the external canthus and never in the whole range of movement. These nystagmoid movements must be differentiated from the violent jerkings seen in the course of a convulsion, these lasting only during the convulsion and always being to the opposite side of the lesion, whereas in the other conditions the jerkings may be to either side, depending on the nature of the lesion.

Prevost³¹ first showed that deviation is toward the lesions when the lesion is cerebral and away from the lesion when it is in the isthmus. Some years later Landouzy¹⁰ advanced the view that irritation and paralysis caused deviation in opposite directions when the lesion was in the cerebrum, the patient looking at the lesion when it is a paralyzing one and at the paralyzed limb when it is of an irritating nature. Grasset²¹ later showed that the opposite was true in lesions of the brain stem. The above facts have been repeatedly demonstrated. It must be remembered, however, that the same lesion may at different times cause paralyzing or irritating symptoms.

Wernicke¹¹ first advanced the view that the rapid disappearance of the conjugate deviation of the head and eyes was owing to the partial control of these movements by the cerebral hemisphere of the sound side and to the action of its oculomotor and head centers. It is also because of the action of the oculomotor center in the unimpaired hemisphere that conjugate deviation can

29. Roussy and Gauckler: *Rev. Neurol.*, 1904, p. 763.

30. Gaussel: *Semaine Médicale*, May 18, 1904.

31. Prevost: *Thèse de Paris*, 1868.

be most readily explained. If one oculomotor center or the fibers in association with it are injured or destroyed they cease to perform their function, at least for a time; therefore, the center in the sound hemisphere causes the deviation to the opposite side, which would cause the eyes to look at the lesion. In irritative lesions as in tumors, this rule is reversed because as in experimental investigations irritation of one side causes movement to the opposite side.

In those cases in which the deviation of the eyes alone, or of the head, occurs, it must be that their respective centers or their associated fibers are separately involved. Similarly, in those rare instances in which the head and the eyes are deviated in separate directions, there must be an irritating lesion acting on one center and a paralyzing one on the other. These clinical facts also argue for separate cortical centers for the eyes and the head.

If we compare the pathologic lesions causing conjugate deviation with others in which no deviation occurs, as, for instance, in a hemorrhage which causes hemiplegia, we may find a most remarkable similarity in their location and extent. Naturally the question arises, why should conjugate deviation occur as a result of one lesion and not of another? As a matter of fact, to a certain extent, this does happen in every case; only in some the symptoms are more marked. The deviation in other words is of minor or partial degree, but nevertheless real. Leichtenstern and Hunnius (cited by Klaas²⁵) have shown that in cases of brain apoplexy in which no deviation occurred there was difficulty in looking to the side opposite the lesion, while the patient could easily look to the side of the lesion. This fact has caused Brissaud and Pechin³³ to call this symptom an "ocular hemiplegia," an excellent term. It may be that conjugate deviation only occurs when the lesion is large and the patient unconscious, or it may argue, according to some, that there is a special tract concerned with conjugate deviation, the existence of which, however, has not been demonstrated.

In 1904 Bard¹ gave a new impetus to the study of conjugate deviation of the head and eyes. He then advanced the view that in the greater number of hemiplegics with conjugate deviation we have present a more or less degree of lateral homonymous hemianopsia, and

33. Brissaud and Pechin: *Rev. Neurol.*, 1904, p. 638.

it is because of an active movement subconsciously commanded by the sensory-motor centers of the sound side that the patient turns the head and eyes of the sound side to regard that which he sees. Bard also says:

The exterior excitations produced by reflexes are not indispensable, the fact that because of the suspension of activities of the one hemisphere, the spontaneous evocation of the sensorial images has not always no part there, is capable of causing the lateral deviation and it is without doubt for this reason that these sometimes persist, just as they may appear during sleep.

Bard also elsewhere says that the sense of sight plays the predominating rôle in conjugate deviation of the eyes and head. His view has been supported by Dufour,²⁹ but denied by Grasset.⁴ The case of Dejerine and Roussy⁵ effectively disproves the views of Bard,¹ for in this patient in whom conjugate deviation occurred there was blindness from birth, showing that there need not be hemianopsia in association with conjugate deviation, contrary to the views of Dufour²⁹ and Grasset,⁴ who believe that hemianopsia is a most frequent symptom. This I also believe to be an error. In most of the cases here recorded it was not possible to demonstrate hemianopsia; in fact, it was only possible to do this in three.

There is still another source of error. In many instances following an apoplectic attack there seems to be what clinically can be more or less satisfactorily demonstrated as a homonymous hemianopsia, but at necropsy the lesion does not involve the optic radiations. This hemianopsia may be transient, and may be demonstrated only for a day or so, or may remain longer, and can be likened to the temporary hemianesthesia so often seen in capsular lesions. In other words, it is my view that many of the hemianopsias demonstrated in these cases are only temporary symptoms and are due either to shock of the hemorrhage or to a fleeting injury of the optic radiations, and that this hemianopsia is the result of the lesion as much as the conjugate deviation and not the cause of the latter.

This is well demonstrated in Case 16. When the patient was examined by Dr. McConnell and myself she had, besides a right hemiplegia, a deviation of the head and eyes to the left. She could look to the left and upward and downward, but could not look to the right past the median line. In testing her with the hand or with the feeding cup a homonymous right, lateral hemi-

anopsia was repeatedly demonstrated. This was not obtained the following day. At necropsy an area of red softening was found involving the anterior portion of the left lenticular nucleus and the caudate nucleus, and a small hemorrhage was found just underneath the left angular gyrus. The optic radiations or the posterior limb of the internal capsule were in no way affected (Fig. 2). The hemianopsia was undoubtedly the result of shock, although the patient was fairly conscious and could obey commands.

To a certain extent hemianopsia does influence the associated movements of the eyeballs, but it probably has no effect whatever in producing conjugate deviation. It is well known that in cases of hemianopsia, whether of peripheral or central origin, the patients have a ten-

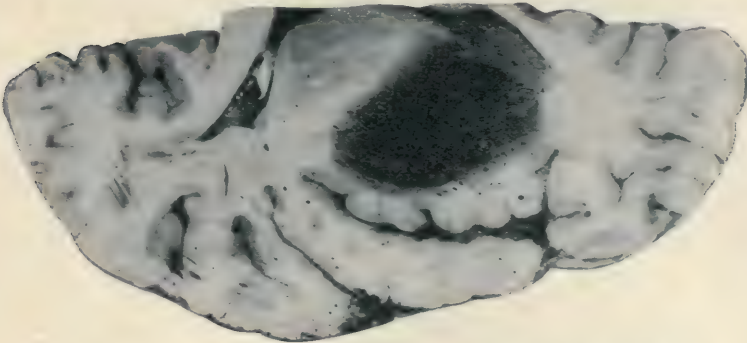


Fig. 2.—Area of softening involving the caudate nucleus and lenticular nucleus, with a fresh hemorrhage underneath the angular gyrus. Clinically, hemiplegia with temporary hemianesthesia and hemianopsia was demonstrated. The two latter symptoms were probably due to the shock of the hemorrhage (Case 16).

dency to deviate their heads, but here, instead of the head being deviated to the side in which one sees, the patient deviates the head to the hemianopic side because he desires to bring into use the part of the retina which is not diseased.

There is still another factor. If a patient has a hemianopsia of any duration he soon learns that he can not look to that side, and in a short time he will accustom himself not to deviate his eyes in that direction. If such a patient were to be suddenly asked to look in the direction of the hemianopsia, he would not be able to deviate his eyes immediately, or to deviate them so promptly as toward the sound side. This I was able to demon-

strate in two cases of hemianopsia due to a basal syphilitic lesion, and in one case was able to judge from this symptom on which side the hemianopsia existed.

In Case 7 paralysis of associated ocular movement existed to the hemianopic side, and this persisted until almost the time of death, for several months. This patient had a large fibrosarcomatous tumor, as shown in photograph (Fig. 1), which occupied the whole of one occipital lobe. This patient had first scintillating scotoma, then hemianopsia, hemianesthesia and hemiplegia. The paralysis of lateral associated ocular movement was not complete, and in this case was probably due to the hemianopsia.

During the active stage of conjugate movement of the eyes the patient can very easily rotate the eyes toward the side of the lesion, and also upward and downward, but in most cases can not deviate them toward the sound side. This paralysis of associated ocular movement is only relative and depends on the conjugate deviation. It is, of course, only temporary.

Irregular or ataxic movements of the eyeballs have been recorded as a result of lesions of the frontal lobe. Thus in a case of Dercum,³⁴ of trauma to the frontal lobe, ataxic movements were present, as were also in a similar case of Klien's.³⁵ These symptoms, however, alone are not diagnostic, but may be valuable when accompanied by other symptoms.

CONCLUSIONS.

The following conclusions can be drawn as a result of the studies in this paper:

1. Conjugate deviation of the eyes and of the head is dependent on a most complex mechanism.

2. In the human being there is but one oculomotor center, or at least one active functioning center, situated in the posterior portion of the second and third frontal convolutions, adjacent to the precentral convolutions.

3. A separate center exists for the movements of the head, probably in the lower anterior portion of the precentral convolution.

4. There is probably in man a distinct center for the combined movements of the eyes and head, situated in the area between the head and the eye centers.

34. Dercum: *Jour. Nervous and Mental Dis.*, Feb., 1905.

35. Klien: *Deut. Ztschr. f. Nervenheilk.*, vol. xxvi, 1904, p. 327.

5. It is probable that the cortical oculomotor, head and combined head and eye centers are subdivided for lateral as well as for upward and downward movements.

6. The oculomotor and the motor head centers are in connection by means of association fibers with the cortical centers for the special senses, in the temporal, occipital, uncinate and other lobes.

7. Any lesion in the motor centers for the eyes and head or in the related special sense centers or in the association fibers connecting the former with the latter will cause an impairment in voluntary deviation of the eyes or of the head, or of both, this depending on the nature and location of the lesion.

8. Lesions in the angular gyrus cause conjugate deviation because of involvement of visual and auditory fibers which lie underneath this area.

9. The theory of Bard¹ that conjugate deviation of the eyes and head is always or nearly always accompanied by homonymous lateral hemianopsia and dependent on this is an error, for in the majority of instances hemianopsia does not exist.

10. Hemianopsia may be caused by the shock or transient effects of the hemorrhage, this loss of half vision being only a temporary symptom and similar in nature to the temporary hemianesthesias sometimes observed in capsular lesions.

11. The occurrence of conjugate deviation of the eyes or of the head, or of both, is of no value as a focalizing symptom, because it may be the result of a lesion in any portion of the cerebrum. It may be of value, however, in conjunction with other localizing symptoms.

12. Conjugate deviation of the eyes and head probably occurs in every case of large apoplectic lesions, but in some instances it is a partial or minor form and of transient duration.

13. Paralysis or impairment of associated ocular movement may occur as a result of a hemianopsia. This, however, is only temporary.

14. There is at present no evidence of a center or centers for automatic ocular movements, but if such centers exist in the thalamus a lesion of these should cause forced or incoördinate ocular movements.

15. Convergence and divergence are probably not reflex acts, but associated ocular movements similar to lateral and upward movements. They probably have

cortical centers in the posterior portion of the second and third frontal convolutions. The movements of convergence and divergence are probably brought about by associating tracts in the pons and cerebral peduncles.

AUTHOR'S CASES OF CONJUGATE DEVIATION OF THE EYE AND HEAD.

CASE 1.—McL., a woman, aged 60, was admitted to the Philadelphia General Hospital, March 13, 1905, to the service of Dr. Spiller from the out-wards, with a history of a stroke of one day's duration, in the left side.

Examination.—The tendon reflexes of the upper limbs seemed to be absent. In the lower limbs the patellar and Achilles jerks were increased, more on the left side, and the Babinski sign was present on this side. Sensation for touch and pin prick was normal. The patient was not wholly unconscious and could respond to questions and would obey commands. A sweetish, urinous odor which suggested a uremic condition was present. The pupils were equal and larger than normal, and the response to light was sluggish on both sides. She could not be tested for accommodation and convergence, and ocular movements because of her mental condition. An occasional tremor was noticed which involved the whole of the left side of the face and the left thumb, this tremor being fine in character. The tremor was at times communicated to the lower part of the right side of the face. Besides, the patient had occasionally a distinct series of convulsive movements which were limited to the left side of the face, the muscles of the face twitching about twice a second, and with this there were twitchings in the muscles of the neck, most marked on the left side, the head being turned to the left with both eyelids closing and opening synchronously with the other contractions. The left thumb was also involved, no movement being apparent in the rest of the hand or of other portions of the body. These convulsions lasted about three minutes, after which the patient would take a long breath as though recovering from an attack of epilepsy.

When examined by me, on the date of death which was five days after stroke, the patient's head was constantly deviated to the left, the eyes deviating to the *right*, there being a constant horizontal movement from the left to the right, the eyes not going past the median line.

Autopsy.—No gross lesions were found in the brain. Two depressions were found on the lateral surface of the left cerebral hemisphere which were filled with fluid, one of these depressions occupying the area of Broca, and extending upward into the lower posterior portion of the second left frontal convolution. The other area was in the lower part of the intraparietal fissure. These areas presented the appearance of a pushing apart of the convolutions without the formation of sclerotic tissue. As they were filled with fluid there must have been pressure exerted on the surrounding brain tissue.

CASE 2.—W., a man, aged 60, had been in the Wills Eye Hospital because of glaucoma, under the care of Dr. S. Lewis Ziegler who referred the case to me. He had been blind for the past three years. While walking down stairs he suddenly became unconscious and fell. He was brought back to the wards and had two convulsions, irregular in character. He persisted in this comatose condition for two days, during which time the head and eyes were deviated to the left, but there was no paralysis noticeable in any of the limbs.

Examination.—When examined by me two days after his fall the head and eyes were strongly deviated to the left, the eyeballs being in a constant to-and-fro lateral movement, the movement being always from the right to the left and never past the median line. There was a distinct weakness of the lower part of the right face and to a less degree of the right upper limb. The right lower limb was very little if at all involved. Sensation seemed normal; the tendon reflexes on the right side were increased and the Babinski response on this side was obtained. Forceful deviation of the head to the right would cause immediate movement of the head back to the left.

Clinical History.—The patient soon became conscious and it was then found that he was motor aphasic, and that he could only answer "Yes, Sir" and the word, "Norah." The paresis of the right limb disappeared almost completely and nothing remained but the weakness in the right lower face, this being also less than it had been at first. The deviation of the head was not so marked, but the deviation of the eyeballs was still as marked as ever. He was then admitted to the University Hospital. Examination seven days after his attack showed almost complete disappearance of the paresis on the right side, including the face. The deviation of the head was entirely absent, but the eyeballs still had a tendency to deviate to the left. Voluntary associated ocular movements were well performed upward, downward and to the left, but to the right he could not bring his eyeballs more than just a little past the median line. Motor aphasia still persisted. It was not until about seventeen days after his attack that he was able to bring his eyeballs completely to the right in associated ocular movements. The motor aphasia was still somewhat present.

Diagnosis.—The patient evidently had a hemorrhage in Broca's convolution on the right side, this involving partially the center for movements of the head, and of the eyes in the adjoining posterior portion of the second frontal convolution.

CASE 3.—McJ., a woman, aged 62, was admitted to the nervous wards of the Philadelphia General Hospital, March 28, 1906, in the service of Dr. Spiller.

Examination.—She was mentally clear and answered questions logically and would obey commands well. She stated that she had had intense headache, some dizziness, nausea and vomiting for the previous three days. While lying passively in bed the head was deviated to the right and she was unable

to turn it to the left. On a forcible attempt to straighten her head the resistance to the movement was very noticeable. It could, however, be forcibly turned completely to the other side, but would immediately turn to the right. The eyes were also deviated constantly to the right. The patient unquestionably had right homonymous hemianopsia, as she was able to recognize all objects properly on her right side but failed utterly on her left. The pupils were unequal and seemed to react slowly to light. Efforts at convergence in looking to the right could only be tested, and when this was done convergence was normal.

During my examination, which lasted about three-fourths of an hour, the patient had five convulsions which would start as follows: There was first a slow turning of the head from the right to the left; at the same time or perhaps a second later there was an upward movement of the left arm as if the hand was held in gesticulation; the head was now held in violent tonic contraction to the left and the face was drawn to the left, the right face drooping; the eyeballs were in constant contraction to the left and were jerked to and fro violently to the left outer canthus. Almost immediately after the movement of the left upper limb the right upper limb was raised and the body was forcibly turned to the left. The tonic contraction of the left hand and face were now succeeded by a clonic contracture of all the limbs of the body. During the tonic contracture the left angle of the mouth was violently drawn upward, the eyelids were alternately opened and closed on both sides, but about three times as rapidly on the left side as on the right. The convulsions sometimes would start in the left side of the face, or again would start simultaneously in the left side of the face and left arm or the right limb. In the majority of instances, however, the convulsion would start with movements of the left face or the left hand.

There was present also a complete left hemiplegia, flaccid in character. The reflexes were increased on both sides, more on the left. The Babinski reflex was present on this side and not on the right. Sensation for all forms was lost completely over the left side. The sense of position was also lost in the left upper and lower limbs and she was unable to recognize any object placed in the left hand.

She was able to look upward and downward with both eyes equally and well. She could look to the right and when this was done the right eyeball was not brought to the outer canthus by one-fourth inch, the left eyeball, however, was fully deviated to the internal canthus. Associated ocular movements to the left were impossible.

Patient died three days after her admission to the hospital.

Autopsy.—Fracture of the cranial bone was found starting at about one-half inch above the ear and extending downward almost to the foramen magnum. The line of another fracture extended from a point between the foramen magnum directly

backward to the right of the lateral sinus. Extensive hemorrhages were found occupying the right frontal pole, this extending in its posterior aspect to the foot of the second frontal convolution and lower part of the precentral convolution. Small hemorrhages were also found involving the right angular gyrus and the right occipital point. Horizontal section of the brain showed the hemorrhage to be cortical in all these points and not involving the white substance in any portion.

CASE 4.—S., adult male, was admitted to Philadelphia General Hospital, Dec. 9, 1902, to the service of Dr. Mills.

Nothing could be obtained of his past history as he was admitted in a stuporous condition and his identity was unknown. A complete left hemiplegia was present. The tendon reflexes were exaggerated, more so on the left side. The Babinski reflex was present on this side. He apparently had a complete left hemiplegia with left hemianesthesia and left hemianopsia. The pupils did not react to light or accommodation on either side. Patient was unable to turn his eyes in associated movements to the right. He died three days after his stroke.

Autopsy.—Several large hemorrhages were found involving the greater portion of the right parietal convolution and extending forward beyond the Rolandic fissure into the back part of the frontal lobe and posteriorly in the occipital lobe.

CASE 5.—B., male, aged 60, a weaver, was admitted to the nervous wards of the Philadelphia General Hospital, in the service of Dr. Spiller, April 4, 1903.

History.—He had syphilis when 30, but otherwise his past history was of no importance. When admitted to the hospital he complained of weakness and stiffness in both lower limbs which he had had for the previous six months, and also some difficulty in talking and swallowing. There was no history of apoplectic attacks.

Examination.—His gait was stiff, his steps were short: station was normal with eyes open and closed; speech was bulbar, it being dull, monotonous and indistinct. He had dribbling of saliva, difficulty in chewing and eating and would even choke. There was no apparent weakness in any of the cranial nerves. The lower portion of the face seemed atrophied, but no tremors were present either here or in the tongue. The upper and lower limbs were weak and spastic, especially the left; all the tendon reflexes on both sides were exaggerated, especially on the left. Ankle clonus was present on both sides and the Babinski reflex was typical on the left and uncertain on the right. Sensation was normal for all forms everywhere. Some atrophy was present on both sides in the thenar and hypothenar eminences, especially on the left side. Some general atrophy was also present.

Diagnosis.—The patient was regarded as a questionable case of bulbar palsy; the slow progress and the absence of more

involvement of the cranial nerves and of fibrillary tremors, making the diagnosis of pseudobulbar palsy possible.

Clinical History.—The condition of the patient gradually became worse, the bulbar symptoms becoming more marked. Two days before his death he suddenly became unconscious and had convulsions, clonic in character, these being limited entirely to the right side. When examined by Dr. Spiller (who made the following observations) the patient was found in an entirely unconscious condition, paying no attention when his name was called; breathing was stertorous and the right cheek was puffed out as though that side of the face was paralyzed. The right upper and lower limb was raised, and dropped like a flail, and irritation of these parts did not elicit any movement. The reflexes were more exaggerated on the right side and this side was more spastic. The Babinski response was prompt on each side. The head and eyes were deviated to the left. If the head was forcibly turned to the right it would soon forcibly turn back to the left. Slight irregular lateral jerkings of the eyeballs were present. The right pupil was larger than the left and the reaction of the iris to the light was slightly present. The deviation of the head and eyes persisted until the following day, when the patient died.

Autopsy.—The case proved to be one of pseudobulbar palsy, as a scar of an old hemorrhage was found in each lenticular nucleus. Besides, a recent hemorrhage was found in the left occipito-temporal region extending to the cortex. This hemorrhage was in the posterior portion of the first and second left temporal convolutions and just below the angular gyrus. The area involved was 4. cm. deep, and 1.2 cm. in its greatest width.

CASE 6.—G., a woman, aged 80, a domestic, was admitted to the nervous wards of the Philadelphia General Hospital, Oct. 12, 1905, in the service of Dr. Mills. Nothing was known either of the family or of past history. She was admitted from the out-wards with a history of suddenly developing numbness of the left hand, followed by pain and convulsions limited to the left face and hand and weakness in these parts.

Examination.—When examined by Dr. McConnell, the patient showed a distinct weakness of the lower part of the left face and of the left upper limb, this weakness in the limb being more marked in the distal parts. The other limbs were not involved. Sensation for touch and pain were normal everywhere. The reflexes in the left upper limb were exaggerated, but the other parts were normal. During the examination the left side of the face and the left hand were the seat of intermittent convulsive movements, the head being drawn somewhat to the left and the left angle of the mouth upward and outward, closing both eyelids and wrinkling the brow, with dilatation of the left nostril. The left platysma muscle was also in contraction. There was no movement of the eyeballs during the convulsion (Dr. McConnell was positive of this, for he observed her especially with this purpose in view). The convulsion

would last for two minutes and did not involve any other portion of the body. The patient was perfectly clear in mentality during the attacks and would answer questions and obey commands directly after the convulsions were over.

Examination of the eyes showed the pupils to be moderately dilated and round, the response to light and accommodation being normal. The left ocular and palpebral conjunctivæ were greatly inflamed. Associated ocular movements upward and to the left were greatly impaired, being in fact almost entirely lost, the loss of upward movement being more marked in the right than in the left eye. The paralysis of associated ocular movements persisted until her death, which occurred four days after her admission to the wards.

Autopsy.—An area of softening was found in the middle of the right precentral gyrus, this softening extending downward as far as and not involving any of the frontal convolutions. A fresh hemorrhage was found in the posterior portion of the occipital cortex, this hemorrhage being 2.5 cm. from the occipital bone. It was 2.5 cm. wide and 2. cm. deep, extending into the white matter and possibly involved the outer part of the optic radiations. The hemorrhage was posterior and inferior to the angular gyrus and very close to it.

Dr. Mills recorded the complete history of this important case before the American Neurological Association, in Boston, June 4 and 5, 1906, in a paper entitled, "Focal Encephalitis."

CASE 7.—McC., a woman, about 50, a patient of Dr. Crothers, of Chester, was seen in consultation by Dr. Mills and myself.

History.—Some notes were obtained from one of her relatives who had been with her constantly for a year or two. This relative stated that the first symptom of which the patient complained were flashes of light which she always saw from the left side. Later the niece observed that she would stumble against objects which were placed on the left, as the left side of the door through which she was passing. This tendency grew worse as time went on and probably indicated a left homonymous hemianopsia. She began to suffer from agonizing headaches, and still later developed nausea and vomiting. It was not until nearly a year after the visual symptoms appeared that she began to develop weakness in the left arm and leg.

Examination.—When she first came under observation patient had a left hemiplegia, the limbs being somewhat contracted. The tendon reflexes were prompt on both sides, but more so on the left, and the Babinski response was present on this side. The patient's mentality was not of the best and it was impossible to tell the state of the sensation. The associated ocular movements were normal to the right and downwards; upward movements were limited, but associated ocular movement to the left was impossible. The pupils were equal and the iritic reflexes were normal.

Eye examination by Dr. C. I. Stiteler showed vision, O. D., 5/9; in O. S., 5/12. The pupils reacted normally, the media

in O. D. was slightly hazy and the outline of the nerve head could clearly be seen only at the upper temporal border, the rest of the nerve being very indistinct and somewhat swollen, especially at the lower border where the margin could not be seen. The retinal vessels were enlarged. In O. S., the media was slightly hazy; the nerve head was normal.

Operation.—There were besides, intense headaches, and some nausea and vomiting. Because of these symptoms of brain tumor an operation was decided on in the motor area. After the operation the patient became considerably better and lived comfortably for two months. In this period homonymous hemianopsia (left lateral) was clearly apparent. Dr. Crothers, under whose care she was, stated that she would never voluntarily look to the left, but that she could look upward, downward and to the right normally; but that towards the last he was able to get her, after violent effort, to look to the right.

Autopsy.—A large tumor was found filling up the whole of the right occipital lobe, extending very nearly to the surface but not breaking into the cortex. The convolutions and fissures over this tumor were much flattened. The tumor extended to the posterior part of the corpus callosum and displaced forward the structures anterior to this. Just at the anterior border of the tumor the wound made by the surgeon at the time of the operation could be found. The angular and supra-marginal gyri were involved in the tumor.

CASE 8.—H., woman, aged 60, a cook, was admitted to the Philadelphia General Hospital, Dec. 15, 1904, in the service of Dr. Mills.

History.—Five days before admission to the hospital the patient suddenly became unconscious and developed a complete right hemiplegia.

Examination.—She was unconscious on admission; the paralysis on the right side was complete and flaccid in character. The reflexes on each side were diminished, but the Babinski reflex was present on the right. The patient soon recovered consciousness and was able to obey commands; sensation was not disturbed and there was no hemianopsia. Motor aphasia was well marked. The following notes were made by Dr. Pemberton, the resident physician: "The right pupil is contracted and apparently immobile to light. The left pupil is dilated and fixed. The media of both eyes is very cloudy, especially the left, which shows a cataract and a marked arcus senilis; accommodation seems to be impossible. The patient looks straight ahead and she cannot apparently move her eyes in any direction when told to do so, although she evidently understands commands and makes an effort to obey, as shown by slight turning of the head."

The patient died nineteen days after the appearance of the apoplectic symptoms.

Autopsy.—A thrombus of the left middle cerebral artery was found, with softening of the brain in the precentral, postcentral

and the whole of the parietal convolutions, that is, in the area supplied by the middle cerebral artery. The softening extended to the lenticular nucleus, but did not invade it, as the thrombus was beyond the point where the arteries to the basilar ganglia arise. The posterior portion of the second frontal convolution was not involved.

CASE 9.—H., a woman, aged 70, was admitted to Philadelphia General Hospital, June 23, 1903, to the service of Dr. Spiller, with a history of right hemiplegia, one day in duration.

Examination.—The paralysis on the right side was complete and flaccid in character. The patient was stuporous and could not be aroused. The reflexes were diminished, this being more so on the paralyzed side. The Babinski reflex was obtained on the right side; not on the left. Pin prick seemed to be perceived on the paralyzed side, as shown by the movements of the opposing limbs. There was decided conjugate deviation of the head and eyes to the left. When the tongue was protruded it was also protruded to the left.

Patient died eight days after her stroke.

Autopsy.—A recent hemorrhage was found in the left optic thalamus, destroying the greater part of the thalamus and extending into the posterior limb of the left internal capsule, but apparently not invading the lenticular nucleus or involving the optic radiations.

CASE 10.—H., a tailor, aged 80, was admitted to Philadelphia General Hospital, March 6, 1906, from the out-wards, to the service of Dr. Spiller.

Examination.—A complete right hemiplegia was present, the paralysis being flaccid in type. The tendon reflexes were increased on the right side and the Babinski response was prompt. On the left side the reflexes were normal. Pin prick was not recognized on the right side, but on the left side any irritation would cause prompt movement. It was impossible to tell whether hemianopsia was present. There was also incontinence of urine and feces. The patient's head was deviated to the left and when it was forcibly turned to the right or to the median line it was immediately turned back to the left. There was also a conjugate deviation of the eyes to the left.

Examination two days later showed practically the same condition, the deviation of the head and eyes being as marked as when examined the first time. There were present besides, constant horizontal movements of the eye, this being perceived with the eyes open, and also when they were shut, the movements being transmitted to the lids. These lateral movements were also to the left. Irregular and jerking movements were also present in the left limbs. Examination two days after this and six days after his stroke, still denoted the same condition, the head not being deviated so strongly. The deviation of the eyes, however, was as marked as ever. Any movements of the head would always be to the left and never to the right, no matter where the irritation was made. The pupils were small and equal; the light reaction was normal. All movements of

the eyeballs were to the left and not to the right. The patient died seven days after his stroke.

Autopsy.—A recent large hemorrhage was found filling up the posterior horn and descending horn of the left lateral ventricle, but not into the anterior horn, and also involving a large part of the lenticular nucleus, doubtless disturbing the function of the left optic radiations and involving also the posterior limb of the internal capsule and the optic thalamus.

CASE 11.—D., a man, aged 52, with a history of syphilis and alcoholism, was admitted to the nervous wards of the Philadelphia General Hospital, Sept. 17, 1902, in the service of Dr. C. S. Potts, to whom I am indebted for the material of this case. A day before his admission the patient had an apoplectic attack, with a resulting left hemiplegia.

Examination.—The paralysis was complete and flaccid, the tendon reflexes were not exaggerated on either side, but the Babinski response was obtained on the left side. A hypesthesia for touch and pain was apparently present in the paralyzed side. The patient had conjugate deviation of the eyes to the right, with inability to turn the eyes to the left.

Autopsy.—An area of softening was found in the lenticular nucleus of the right side, and extending into the posterior limb of the internal capsule, but not invading the optic radiations.

CASE 12.—R., a man, aged 77, was admitted to the nervous wards of the Philadelphia General Hospital, July 19, 1904, in the service of Dr. Spiller.

History.—A sudden attack of unconsciousness the day before, resulting in a left hemiplegia.

Examination.—On the following day examination showed a complete left hemiplegia, the paralysis being flaccid. The tendon reflexes were lost on the paralyzed side and were diminished on the sound side. The Babinski response was obtained on the left side. Pin prick was not recognized at any portion of the left side, but was promptly recognized on the right side. The patient was not totally unconscious, as he would obey commands, as sticking out his tongue and lifting up his limbs of his sound side. There was also present incontinence of urine and feces. The eyes were distinctly deviated to the right, but there was no deviation of the head. It was impossible to tell if hemianopsia was present.

The patient died six days after his stroke.

Autopsy.—A large hemorrhage was found destroying the posterior portion of the right lenticular nucleus, extending backward beyond the nucleus almost to the angular gyrus and destroying the posterior part of the posterior limb of the internal capsule. This large hemorrhage did not involve the cortex at any point, although in one part minute hemorrhages were found in a portion of the cortex about the island of Reil.

CASE 13.—S., a man, aged 51, was admitted to the Philadelphia General Hospital, Sept. 21, 1903, into the service of Dr. Mills.

History.—He had been admitted to the hospital several years previously with a history of right hemiplegia, from which he partially recovered.

After being in the hospital for two years he suddenly had an attack of unconsciousness. The resident physician, Dr. Maier, found him in a stuporous condition, the right arm and leg being completely paralyzed and the right face drooping. Some spasticity was present on this side and the reflexes were exaggerated. The head was not deviated to either side; the eyes were turned up and oscillated slightly from the right to the left. He died two days afterwards.

Autopsy.—A lesion was found in the left lenticular nucleus not extending apparently into the internal capsule, and reaching and invading the claustrum, extreme capsule and not reaching the cortex. The lesion must have involved the fibers underneath the angular gyrus.

CASE 14.—A. was an old right hemiplegic, who had for a long time been in the nervous wards of the University Hospital. His previous records were lost.

One morning he suddenly became unconscious and had convulsions, which were limited to the left upper and lower limbs and the left side of the face. The eyes were turned in conjugate deviation to the right. Both upper and lower limbs were flaccid, there being, however, more tonicity in the left side. The tendon reflexes were lost in all the limbs, but the Babinski response was distinct on the right side. The patient died the same day.

Autopsy.—The ventricles were found to be considerably dilated and the scar of an old cyst was found in the external portion of the left lenticular nucleus and involving the extreme capsule.

CASE 15.—C., a woman, was admitted to the nervous wards of the Philadelphia General Hospital, July 12, 1904, into the service of Dr. Spiller.

History.—The patient then had a paresis on the left side of the body. Feb. 22, 1905, she suddenly became paralyzed, about seven months after her first stroke.

At this time Dr. Spiller made the following dictation: "The patient is in a deep stupor and is unable to understand anything said to her. The eyeballs are deviated to the right and there are frequent irregular jerky movements to the right, the left edge of the cornea going but very little beyond the median point of the palpebral fissure during these movements. These clonic movements of the eyeballs are suggestive of an irritation of the left cortical center of the ocular muscles."

When she was seen by Dr. Pemberton, the resident physician, somewhat previously, but on the same day, the eyes were turned to the left, and there were no jerky movements of the eyeballs. She had therefore conjugate deviation of the eyes to the left, but this was at the time when there was no irritation of the eye muscles, and therefore would seem to indicate

that she was looking at her lesion. The head was not stiff and remained where placed; there was also a slight tendency of the eyeballs to be drawn upward as well as to the right. The pupils were dilated, and equally so, and did not respond to light. Occasionally an involuntary jerking of the left upper limb was observed. Both upper and lower limbs were flexed and the reflexes were diminished, the left more than the right. Babinski response was present on the left but not on the right. She died the following day.

Diagnosis.—Left hemiplegia.

Autopsy.—A limited area of sclerosis of the anterior portion of the right second frontal convolution was noted. There was also a cyst in the anterior portion of the left lenticular nucleus. An area of softening in the right frontal lobe at the frontal point was found. The lateral ventricles were also somewhat dilated.

CASE 16.—C., a woman, aged 35, was admitted to the Philadelphia General Hospital, Dec. 1, 1905, to the service of Dr. Hirst.

History.—Patient was delivered of a child four weeks previously, since which time she had been having a temperature and had apparently been septic. Two days before her admission she suddenly fell, became unconscious, and has not been able to speak since.

Examination.—She was examined five days after her admission by Dr. McConnell and myself. The patient then had a complete right hemiplegia, the limbs on the paralyzed side being spastic and contracted in the usual hemiplegic position, the reflexes were exaggerated on both sides, the right more than the left, and the Babinski response was present on this side. The patient was not unconscious and would obey commands. The head and eyes had been deviated since her admission constantly to the left. Pin prick or touch on the right side could not be recognized anywhere, but on the left side sensation was promptly recognized. Objects were not recognized in the right visual fields of either eye, as was demonstrated by the feeding cup and other tests, therefore, right homonymous hemianopsia was present. The patient was aphasic, but could understand questions and would nod responses. The right masseter muscle was in a constant state of contraction, causing the right jaw to be locked. She was able to turn her eyeballs upward, downward, and to the left, but could not turn them to the right. When examined a day after this the patient could turn her eyes a little to the right, but not normally so. She died a few days afterwards as the result of a double pneumonia.

Autopsy.—An area of red softening was found, involving the left caudate and the left lenticular nucleus. A small hemorrhage was also found in the cortex of the upper portion of the first temporal convolution just below and anterior to the angular gyrus.

1

PATHOLOGY OF PARALYSIS AGITANS, WITH
A REPORT OF FOURTEEN CASES
WITH NECROPSY.*

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PHILADELPHIA.

The subject of the pathology of paralysis agitans is one on which few physicians have definite ideas, and those who have differ widely from each other. It is obvious that so long as the nature of this disease is in controversy and obscurity the treatment must be imperfect, changeable and unfortunately inefficient. Numerous investigations with the idea of clearing up this problem have but added to its complexity by suggesting new hypotheses and unexplored fields for reasearch.

The clinical picture of paralysis agitans is so clear cut and typical, the course of the disease so unchangeable and progressive that it certainly suggests an organic basis. To classify it as a neurosis is begging the question and explains nothing, while it would be distinctly harmful if it hindered further search for the real pathology.

SUMMARY OF LITERATURE.

In order to define the present status of our knowledge of the subject, it will be necessary to review briefly the findings and opinions recorded by numerous observers since Parkinson wrote his classical paper in 1817. For the sake of convenience these findings may be classified

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according to their location into: 1. Those in the nervous system, the brain, spinal cord and peripheral nerves. 2. Those in the muscles. 3. Those in the ductless glands.

The gross lesions in the central nervous system found by some of the earlier writers which were regarded by them as the cause of paralysis agitans are not now credited with such a relationship. It must be remembered that at that time the difference between multiple sclerosis and paralysis agitans was not well understood; consequently, in the opinion of Wollenberg,¹ one of the cases which Parkinson himself described was probably a case of multiple sclerosis and, according to the same authority, similar cases have been reported by Oppolzer,² Marshall-Hall,³ Skoda,⁴ Lebert⁵ and others.

Confusion has arisen, also, from the fact that cases are reported in which paralysis agitans was combined with multiple sclerosis, as in the case reported by Furstner⁶ (his second case) and the case of Schultz,⁷ in which the diagnosis is questionable, as he only found areas of sclerosis of pinhead size ("Stecknadelkopfgrosse"), but Sanders⁸ thinks it should be included in this category. Many of the earlier writers recorded as paralysis agitans cases in which the tremor was symptomatic of some gross lesion of the brain, such as the case reported by v. Leyden⁹ of a sarcoma of the left optic thalamus, also those of Charcot,¹ Benedikt,¹ and Blocc and Marinesco,¹⁰ who described cases of tremor caused by tumors situated in the region of the cerebral peduncles. Chvostek¹¹ reported a case in which clinically there was first paresis, then tremor of the extremities on the left side, and in which anatomically there was a sclerosis of the entire right cornu ammonis. Ordenstein¹² found in one case areas of softening in the cerebral peduncles, but in the two other cases reported by

1. Nothnagel: spec. Path. u. Therap., xii.

2. Wien. med. Wochschr., 1861, Nos. 36-38. Quoted by Wollenberg, reference 1.

3. "Traité des Maladies du Systeme Nerveux," 1841.

4. Wien. med. Halle, 1862, No. 13.

5. "Handb. d. prakt. Med.," 1871, ii, p. 632.

6. Arch. f. Psych. u. Nervenkrankh., 1898, xxx.

7. Virchow's Arch., lxxviii, 1876, p. 120.

8. Article by Sanders: Monatschr. f. Psych. u. Neurol., 1898.

9. Virchow's Arch., xxix, p. 202, 1864.

10. Ref. in Neurol. Centrbl., 1894.

11. Wien. med. Wochschr., 1871, Nos. 37 to 39.

12. Thèse de Paris, 1868.

him at the same time the findings were negative. Herterich¹³ described a case with very marked contractures in which he found an area of softening in the left cerebral peduncle. Cases have been reported by Virchow,¹⁴ Leubuscher,⁸ Berger⁷ and others in which areas of softening in the brain were the cause of a tremor. In reporting a case of tumor of the left prefrontal lobe, Mills and Weisenburg¹⁵ say:

There were muscular tremors in the hands and legs much like those seen in paralysis agitans. At first they were on the left side only, but toward the end involved the right side also. They were at first partially under control, and by an effort of will the patient could stop them; later he could not, and they occurred also during sleep.

Though this tremor differed in some respects from the tremor of paralysis agitans, it is difficult to see why, if a symptomatic tremor, it should have begun on the same side as the tumor. There are many instances of gross lesions found in the brain which are simply conditions complicating the paralysis agitans, viz.: Oppolzer¹⁶ found an apoplectic cyst in the occipital lobe accompanied by an ependymitis of the lateral ventricles, especially the posterior horns; v. Sass¹⁷ reported a case in which there was an ependymitis of the fourth ventricle. At present the occurrence of gross lesions in the central nervous system in cases of paralysis agitans is regarded as merely a coincidence.

A critical study of the positive pathologic findings in the central nervous system in cases of paralysis agitans reported by numerous investigators leads to the conclusion that they are simply the changes of senility modified occasionally by intercurrent affections or the senile marasmus that is so often the cause of death in these cases. The lesion regarded as specific for the disease by one observer is expressly denied by many others. In most of the cases the lesions described are diffuse and affect various anatomic elements, nerve fibers, nerve cells, neuroglia, the blood vessels, etc., but there is the greatest diversity of opinion as to which is the most affected,

13. Herterich Dissert., Würzburg, 1878. Quoted by Keller, reference 16.

14. Virchow: Die krankhaften Geschwülste, xl. Cited by Sanders, reference, 8.

15. THE JOURNAL A. M. A., 1906, No. 5.

16. Article by Koller: Virchow's Arch., cxxv, p. 287.

17. Petersburg Wochschr., 1891, Nos. 19-20, abs. in the Neurol. Centrabl., x, p. 756.

and it is evident that these findings have no constant relation to the disease. Occlusion of the central canal by the overgrowth of the epithelial cells is described by Joffroy,¹⁸ Demange¹⁹ and many others. In the case reported by Murchison and Cayley²⁰ the central canal was widened and filled with leucocytes. In many cases numerous amyloid bodies were found in the spinal cord. Cases are reported as presenting a partial degeneration of the tracts in the white matter of the spinal cord. Burzio²¹ found a light grade of atrophy in many fibers of the crossed pyramidal tracts and of Goll's and Burdach's columns. Dowse²² found degeneration of only one lateral column. Sanders⁸ found "a somewhat diffuse degeneration of nerve fibers in the spinal cord, but especially in the direct and crossed pyramidal tracts." Most investigators expressly deny the existence of any degeneration in the spinal cord, and in the 14 cases studied by me I was unable to detect any degeneration in the Marchi methods of staining.

Many writers have described changes in the neuroglia in paralysis agitans. Schultze speaks of a chronic "leptomeningitis and chronic interstitial myelitis." Murchison and Chayley found thickening of the subpial neuroglia layer with extension of the proliferation into the cord and islets of sclerosis scattered throughout the spinal cord by either the Weigert hematoxylin or the cord, especially in the dorsolumbar region and more in the posterior root entrance zone. Demange,¹⁹ Joffroy¹⁸ and Dubief²³ report similar findings. Koller¹⁶ found practically the same in his case, but questions whether we can draw a line between this proliferation and the neuroglia increase in senility. In paralysis agitans the neuroglia overgrowth, he thinks, was more marked in the lateral and posterior columns, and he considers it to be the primary change, which by interfering with the lymph circulation causes secondary alteration of the nervous elements. Redlich²⁴ lays great stress on the increase of glia tissue which he found, especially about

18. *Gaz. de Hôp.*, 1871, p. 602.

19. *Rev. Méd. de l'Est*, 1897.

20. *Trans. Path. Soc., Lond.*, 1871.

21. *Riv. spermin di frenatria*, 1903, No. 4, 1904, No. 1, abs. in *Centrabl. f. allg. Path. u. path. Anat.*, xvi, p. 700.

22. *Trans. Path. Soc., Lond.*, 1879, xxix, p. 17.

23. *Thèse de Paris*, 1887.

24. *Jahrb. f. Psych.*, xii, p. 385.

the blood vessels, forming small islets which had a tendency to become confluent and form larger islets. This perivascular sclerosis occurred chiefly in the anterolateral columns, and the stiffness of the muscles and the tremor, in his opinion, are due to a functional disability ("schwäche") of the nerve fibres. Somewhat similar findings are described by Wollenberg,¹ Borgherini²⁵ and Sanders,⁸ but the last two mentioned found the neuroglia proliferation more marked in the gray matter than in the white. Sanders especially emphasizes this as being a different condition than that found in senility. He found not only a comparatively greater increase in the amount of neuroglia tissue in the anterior horns, but also numerous spindle cells ("spinnenzellen") in both cases of paralysis agitans and cases of senility with a marked tremor. He explains the tremor and also the difficulties in gait by assuming that this glia overgrowth interferes with the connection between the upper and lower motor neuron. As contrasted with these findings, Dana²⁶ describes a very moderate increase in the connective tissue within the cord and none of the islets of neuroglia described by Redlich.²⁴

Pathologic changes in the blood vessels of the central nervous system are described by many writers on the subject. They are generally of the arteriosclerotic type, but in some cases are described as peculiar either in character or distribution. Burzio²¹ found the blood vessel walls thickened, especially in the neighborhood of the neuroglia overgrowth, and Sanders emphasizes this relation. Sanders found also that the finer capillaries were more affected than the larger, a fact which has been noted by Joffroy,¹⁸ v. Sass¹⁷ and by Murchison and Cayley.²⁰ The last named and also Ketscher²⁷ found miliary hemorrhages here and there in the spinal cord. J. R. Hunt²⁸ found that the blood vessels in the spinal cord were increased in number and there was a universal cellular infiltration about them which, when stained by Erlich's method, proved to consist of escaped leucocytes. Miliary aneurisms were seen in the spinal cord by Borgherini,²⁵ Ketscher²⁷ and Sanders.⁸ Carrayou²⁹

25. Article by Borgherini: *Wien. med. Wochschr.*, 1890.

26. *Am. Jour. Med. Sci.*, November, 1899.

27. *Prager Ztschr. f. Heilk.*, December, 1892.

28. *Jour. Nerv. and Ment. Dis.*, 1896.

29. *Thèse de Paris*, 1903. Abs. in *Le Sémaine Méd.*, 1903, No. 16.

found varicosities of the veins in the medulla with small areas of softening in the oculomotor root and pyramidal tracts, and he explains the clinical symptoms of the disease on the ground that these lesions interfere with the passage of the nervous impulses. Redlich²⁴ speaks of a hyaline degeneration of the blood vessel wall with an especial thickening of the media and a proliferation of tissue about the blood vessel which fills up the perivascular spaces. These changes, he affirms, were much less marked in the medulla oblongata and were not there accompanied by the perivascular sclerosis. Dana²⁶ could discover no special thickening of the walls of either the veins or arteries in his case. Borgherini²⁵ found changes in the sympathetic ganglia, degeneration of the cells with an overgrowth of the interstitial tissue.

Philip³⁰ found degenerated Betz cells in the paracentral lobules. The change consisted in a diffuse staining of the cells and a similar though less marked condition was noted in the motor cells of the anterior horns of the spinal cord. His finding is the only direct pathologic evidence I could discover bearing out the oft repeated assertion (Gowers,³¹ Bychowski³² and others) that paralysis agitans is a disease of the cerebral cortex. Gordinier³³ found changes in the motor cells of the cerebral cortex, but not so marked there as in the spinal cord. The Golgi stain showed the dendritic processes nearly normal.

The condition of the anterior horn cells of the spinal cord has been noted as normal in some of the cases to which reference has already been made (Redlich,²⁴ Wollenberg¹ and others). In a large number of cases the anterior horn cells were found to contain an excessive amount of yellow pigment (Dubief,²³ Dowse,²² v. Sass,¹⁷ Hunt,²⁸ Borgherini,²⁵ Herterich,¹³ Joffroy,¹⁶ Ketcher,²⁷ Gordinier,³³ Luys³⁴ (of the medulla oblongata), Sanders⁸ and others. Joffroy³⁵ and Demange¹⁹ describe also a pigmented condition in the cells of the column of Clarke. Gordinier³³ also noted this, but found it less marked than in the anterior horn cells. Distinct degenerative changes in the anterior horn cells are described by

30. Deutsch. Ztschr. f. Nervenhlk., 1899, Nos. 5-6.

31. Allbutt's "System of Medicine," ix, p. 73, 1900.

32. Arch. f. Psych. u. Nervenkrankhl, 1898, p. 722.

33. Am. Jour. Med. Sci., 1899, p. 648.

34. Gaz. Med. de Paris, 1880.

35. Arch. de Physiol., 1872, No. 1.

Demange,¹⁰ Koller,¹⁶ F. Burzio,²¹ Caterina,³⁶ Hunt,²⁸ Gordinier³³ and Dana.²⁸ The changes described as present in these cases differed somewhat, but were generally a chromatolysis, with a loss or displacement of the nucleus of the cell, atrophy of the cell, vacuolation of the cell, tumefaction of the nucleolus, etc. Dana²⁶ lays great stress on diminution in number and pathologic changes in the dendritic processes of the spinal motor cells, with a consequent diminution in richness of the dendritic plexus in the anterior horns. He found this lesion most marked in the lower cervical and first dorsal segments (corresponding to the beginning of the tremor in his case), next in the midcervical, then midlumbar, upper sacral and least in the middle and upper dorsal. The brain cortex showed slight degeneration of the cells. It should be remembered that Dana's case was complicated by sarcomatosis, and death was due apparently to exhaustion, and that in such cases a tumor toxin is probably present which produces chromatolytic and degenerative changes in the nerve cells.

No pathologic changes in the cerebellum were noted by Dowse,²² v. Sass,¹⁷ Borgherini,²⁵ Redlich,²⁴ Hunt,²⁸ Gordinier³³ or Dana,²⁶ except participation to a moderate extent in a general arteriosclerosis. Hunt²⁸ and Gordinier³³ found the spinal nerve roots normal. Dana²⁶ found them greatly congested, and by Borgherini²⁵ and Redlich²⁴ they were found slightly degenerated.

A long list could be made of cases in which the pathologic examination of the central nervous system was negative. Thus Koller¹⁶ cites the cases of Peträus,³⁷ Olliver,¹⁶ Cohn,³⁸ Charcot,³⁹ 3 cases; Westphal,⁴⁰ Simon,¹⁶ 4 cases; Bauer,⁴¹ 2 cases; Leroux⁴² and Weinkowitz.⁴³ To these may be added the case of Oppenheim,⁸ in which he found only a "questionable" staining of the columns of Goll. Furstner⁶ mentions specifically that there was no pathologic condition present in the brain.

36. Riv. di path. nerve. ment., 1899.

37. Den Fremadskridende Skjoelvelamhed, Hosp. tid. kjobenh., 1861, cited by Koller, reference, 16.

38. Wien. med. Wochschr., 1860, p. 406.

39. "Lecons sur le Systeme Nerv.," Paris, 1884.

40. "Charite-Annalen," 1876, p. 360.

41. Annalen d. stadt. allg. Krankenh. in Munchen, I, p. 124.

42. Diss. Paris, 1880.

43. Diss. Breslau, 1883.

or spinal cord of his first case, the blood vessel walls were not thickened and there was no increase of the neuroglia nor changes in the nerve cells. His patient was 61 years of age and had the disease for five years. Schwenn⁴⁴ reported a case of a patient who died at the age of 43½ years and was therefore no senile. He says: "I found neither in the brain nor spinal cord any pathologic changes." Idelsohn⁴⁵ and Blocq⁴⁶ also report cases in which there was no pathologic change in the central nervous system.

Various changes have been observed in the peripheral nerves. Borgherini²⁵ found in the vagus, median and external tibial nerves a hyperplasia of the interstitial tissue and increased vascularity. Skoda⁴ found thickening of the neurolemma in the nerves of the upper extremities. Joffroy reported a case in which paralysis agitans was combined with a peripheral neuritis, and Follet and Marquis⁴⁷ report *Un cas de maladie de Parkinson fruste, peut-être d'origine polyneuritique*. Their paper is inaccessible to me. Borgherini,²⁵ Redlich,²⁴ v. Leyden⁷ and Ketscher²⁷ have noted an increased amount of the interstitial tissue in the nerves, and by the last-named the nerves were also found degenerated. Blocq,⁴⁶ who examined the peripheral nerves, both in the fresh state and after hardening, found nothing abnormal. Dana²⁶ also found them normal. v. Sass¹⁷ found the perineurium and endoneurium thickened, and they appeared partially degenerated when stained by the method of Pal.

ORIGINAL INVESTIGATIONS: THE NERVOUS SYSTEM.

I have had the opportunity of examining the pathologic material from 14 cases of paralysis agitans, in 8 of which I was able to study the peripheral nerves and muscles and in 2 of them also the ductless glands. That 14 cases with necropsy is a very large number will be better appreciated when it is remembered that Gordinier,³³ writing in 1899, could find reported but 54 cases with necropsy in the whole literature of the disease, and of these only 24 had been examined histologically. The largest number of cases with necropsy reported by any

44. Deutsch Arch. f. klin. Med., 1901, p. 193.

45. Rev. Neurolog., 1904, p. 394.

46. Manuel de Med., Debove Archard, 1894. Quoted by Raymond, reference 57.

47. Bull. Soc. Sci. et Med. de l'Ouest., xl, 104.

single observer is 4. Details of the positive pathologic findings in 14 cases with necropsy, in 12 of which there is also given an abstract of the clinical histories, will be found in the appendix.

The ages of the patients at death, in the cases that I have examined, ranged from 45 to 75 years; the duration of the disease from two to twenty years.

Macroscopic examination of the central nervous system, including trans-sections, revealed lesions in only one case.

The tissues are preserved in formaldehyd solution (10 per cent.), or in Müller's fluid. In making the microscopic examination of the central nervous system pieces of tissue were taken from each paracentral lobule, the pons, the vermis of the cerebellum, the medulla oblongata and from the cervical, thoracic and lumbar regions of the spinal cord. The anterior and posterior spinal nerve roots from the lumbar and cervical regions were examined separately from the cord in 6 cases; the posterior root ganglia, from the lumbar region, in 2 cases; the peripheral nerves were examined in 8 cases. The pieces were dehydrated in alcohol in the usual manner, imbedded in celloidin, and the sections stained as follows: Thionin modification of the Nissl method for study of the nerve cells; the Weigert hematoxylin and the Marchi osmic acid for degeneration; the hemalum-acid fuchsin, and the Mallory neuroglia stains.

A summary of the findings in my cases shows that the most constant lesion was a fibrosis of the capillary blood vessels of the spinal cord, which by rendering them more prominent, caused an apparent increase in their number. The region of the posterior and lateral columns seemed to be the most affected. This lesion was present in eleven cases, in all of which it was an accompaniment of a general sclerosis of the blood vessels of the nervous system. The next most frequent finding was a diffuse overgrowth of the glia in the spinal cord, usually more marked in the posterior columns, in six cases. It was not confined to any system of fibers, and in but one case did it show the tendency to form islets about the blood vessels as described by Redlich.²⁴ In no case was it demonstrated that the neuroglia proliferation was more marked in the gray matter than in the white matter. The Betz cells of the paracentral lobules were considerably pigmented in many cases, but only in four cases was the

pigmentation very marked, and in only two cases were there any distinctly degenerated cells. The motor cells of the anterior horns of the spinal cord were excessively pigmented in six cases; in four cases they showed distinct chromatolytic changes in some of the cells, but the loss of the dendritic processes, with a consequent lessening of the richness of the dendritic plexus, as described by Dana, was not observed. Recent hemorrhages into the anterior horns of the spinal cord were observed in two cases; they were probably agonal. The perivascular spaces were widened in four cases, and in five cases numerous amyloid bodies were seen in the spinal cord, especially in the posterior columns. The central canal was closed in eight cases, dilated in one case, double in one case, but probably these are to be regarded as normal conditions. The cerebellum, pons and medulla oblongata were practically normal in all the cases in which they were examined; a moderate sclerosis of the blood vessels and a slight subpial proliferation of the neuroglia not being considered pathologic when found in aged persons. In one case there was a round cell infiltration of the pia. The anterior and posterior spinal nerve roots in the cervical and lumbar regions were found congested in two cases, but not in any case degenerated. There was no degeneration of the nerve fibers of the cord in any case, but the neuroglia proliferation, in six cases, gave the posterior columns the appearance of staining lighter than the rest of the cord by the Weigert hematoxylin stain. The peripheral nerves were examined in eight cases; in two they appeared degenerated when stained by the Weigert hematoxylin stain; in two others there was a swelling of the myelin sheaths, accompanied in one case with swelling of the axis cylinders. The intramuscular nerves, even those within the muscle spindles, stained well with the Weigert hematoxylin stain. The nerves showed no degeneration when stained by the Marchi method.

DISCUSSION OF THEORIES OF THE NERVOUS ORIGIN OF THE DISEASE.

Many theories have been advanced as to the nature and location of a lesion in the central nervous system in paralysis agitans. Some have been purely hypothetical and based entirely on the clinical symptoms, others on

the findings in the few cases examined by their authors. Gowers³¹ expresses his view thus:

Paralysis agitans must be regarded as a chronic senile change in the nutrition of the terminations of the branching processes of the dendrites of the motor nerve cells of the cortex causing constant liberation of nerve energy to be increased, and also causing a change in the form, uniformity gives place to rhythmical liberation.

Bychowski³² and others hold similar views. The arguments advanced in support of this are: 1. A hemiplegic distribution of the tremor speaks for a cortical origin. Numerous cases have been reported in which the symptoms involved both arms or both legs, or in other words, were paraplegic. By Hart (l. c.) they were found to be as frequently paraplegic as hemiplegic. 2. The tremor ceases during sleep, i. e., when the function of the hemisphere is least active, but in cases in which tremor is known to be due to cortical involvement (ease of Mills and Weisenburg, l. c.) the tremor does not disappear during sleep. 3. When a case of paralysis agitans in which all four extremities are affected develops a hemiplegia, the tremor disappears on the paralyzed side. Such cases are said to have been observed by Gowers,³¹ Westphal⁴⁰ and Grashay.⁴⁸ This, if invariably true, proves that the nervous impulses which by their perversion cause the symptom originate in the cerebral cortex, but it does not prove where the perversion of impulse in this disease takes place. Whether in origin, transmission or in their reception ultimately by the muscle, the result of a complete cessation of the impulse would be the same. Hart reports a case in which the tremor did not stop on the paralyzed side after a hemiplegia and was apparently unaffected by it.

Bychowski³² adds the following reasons: 4. The not uncommon combination of insanity, hysteria or brain disease with paralysis agitans. Most authors regard this combination as decidedly uncommon. 5. The parasthesiæ and subjective sensations of heat and cold. These are generally considered as accompanying peripheral rather than cortical conditions. 6. The constant failure of any change in the sphincter reflexes. This is of some importance as differentiating a spinal from a cortical origin, but would have no bearing if the lesion is outside

48. Berlin klin. Wochschr., No. 30, 1885, p. 487.

the nervous system. 7. The increased knee jerks. The knee jerks in paralysis agitans are too variable to be used as an argument on which to base a pathology of the disease.

Cuylits⁴⁹ considers the tremor of paralysis agitans as cortical in origin, and holds that the lesion consists of retraction of the dendritic processes of the cells and draws analogies to tremor seen in paresis, alcoholism, etc. There is no pathologic evidence to support the theory of a cortical origin for the disease except the findings of Philip.⁵⁰ Brissaud⁵⁰ believes that a lesion of the locus niger is the cause of the disease. Tremors are produced by lesions in this region, but they are different from that of paralysis agitans and are not accompanied by the other symptoms of the disease. Valery⁵¹ considers that paralysis agitans is a syndrome due to a lesion in the mesencephalon, the syndrome being more or less complete, according to the extent of the lesion. However, lesions of the mesencephalon are not found at necropsy.

Ballet⁵² and also Dana,²⁶ from pathologic evidence furnished by their own cases, place the chief lesion in the motor cells in the anterior horns of the spinal cord. Ballet,⁵¹ as a breaking of the processes, particularly the dendritic processes. Dana²⁶ holds that:

There is early functional disturbance and later a degeneration and destruction of the dendrites of the anterior horn cells which interferes with the even flow of motor impulses, and finally leads to motor weakness and rigidity, owing to the cell being practically cut off from the brain.

From a pathologic standpoint this theory is not confirmed by any other observer, although especially looked for in my cases. Clinically, it would seem that a tremor due to such a lesion, "which interferes with the even flow of motor impulses," would be rather of the intention type, and unlike that of paralysis agitans which momentarily disappears on voluntary movement.

Redlich²⁴ believes that the overgrowth of the neuroglia in islets about the blood vessels in the lateral and posterior columns of the spinal cord accounts for the tremor and rigidity on the grounds of a functional weak-

49. *Préssé Méd. Belge*, Brussels, 1897, xlix, p. 121-124.

50. *Jour. de Méd. et de Chir. Pratique*, 1894, p. 641.

51. *Thèse de Paris*, 1904, Abs. *Rev. Neurol.*, 1905.

52. *Bull. et Mem. Soc. Méd. de Hôp. de Paris*, 1898, xv, pp. 40 to 43.

ening of the nerve fibers in their vicinity, thus producing a special type of dorsolateral sclerosis. The rigidity of paralysis agitans, however, is very different from the spasticity due to lesion of the lateral column, and the pathologic findings of Redlich are not confirmed; indeed, are expressly denied by other observers (Dana²⁶). I found a diffuse overgrowth of the neuroglia in the lateral and posterior columns in a number of cases, but not in the form of islets about the blood vessels except in one case.

Gordinier³³ explains the tremor and rigidity as follows:

The characteristic symptoms of this disease are due to one and the same cause, namely, an irritation from the presence of an increased amount of neuroglia in the posterior columns producing an excitability of the reflex collaterals of the posterior nerve roots arborizing about the ventral cornual cells which results in an increase in the normal rhythmic discharges of energy from those cells, producing at first the tremor and later the rigidity.

Gordinier's case, the pathology of which is the basis of his hypothesis as to the causation of tremor and rigidity, was "a typical case without tremor."

Sander⁸ thinks that the overgrowth of glia in the anterior horns by interfering with the connection between the upper and lower motor neuron, is the cause of the tremor and the difficulties in gait. He examined the central nervous system from cases of senility with and without tremor and also paralysis agitans; in the cases of paralysis agitans and in those seniles in whom tremor had been marked, he found a special predilection of the glia proliferation for the gray matter. This theory of Sander⁸ takes no account of the clinical difference between paralysis agitans and senile tremor, and his pathologic findings lack confirmation.

From the above summary it will be seen how diverse and contradictory are the pathologic findings in the central nervous system in paralysis agitans, but the explanation for this is simple. Paralysis agitans is a disease that in almost every case begins late in life and is not rapidly fatal. There are changes which occur in the central nervous system, as they do in other parts of the body with the advance of years, changes that are well known, though perhaps not well understood, and which consist, to state it in a few words, of a gradual replace-

ment of the parenchymatous structure by the connective tissue of the part, in the nervous system by the neuroglia. This change is modified, advanced or retarded, to an extent, by the mode of life, heredity, disease, etc. Arteriosclerosis is usually found at this period of life, and also the incidence of wasting diseases and malignant new growths, which, by their toxic influence, are known to produce degenerative changes in the central nervous system. These changes, the changes of senility variously modified by the other factors of which I have spoken, are the most constant findings in the central nervous system in paralysis agitans.

Many men (Wollenberg,¹ Campbell,⁵³ Nonne⁵⁴ and others) have found changes similar to those found in paralysis agitans, in the spinal cords of seniles who during life had shown none of the symptoms of that disease. To reconcile this similarity in the findings with such distinct clinical differences, it has been held that the lesions in paralysis agitans are of precocious onset and exaggerated intensity; such a view is held by Dubief,²³ Jacobsohn,⁵⁵ Borgherini²⁵ and Ketscher.²⁷ Against this, however, are the findings of Furstner,⁶ who found no marked senile changes, Idelsohn,⁴⁵ and Schwenn,⁴⁴ whose patient died at 43½ years, and was therefore not a senile.

That paralysis agitans is but senility, even though a precocious or special form of senility, I believe, with Dana,²⁶ "is contrary to all clinical experience." The lesions of senility found in the central nervous system in many cases are but natural when we consider the age of the patient at necropsy, and the same lesions are found in many seniles not the subject of the disease; while in those patients who die at an earlier age these changes are not found. Therefore, from a pathologic standpoint also, paralysis agitans and senility must be regarded as two different conditions.

THE CHANGES IN THE MUSCLES.

Oppenheim⁵⁶ gives as the characteristic symptoms of this disease: tremor, muscular rigidity, which produces a peculiar position of the head, trunk and extremities, re-

53. Jour. Ment. Sci., Oct., 1894.

54. Deutsch. Ztschr. f. Nervenheilk., xiv, p. 193, 1899.

55. Berlin klin. Wochschr., No. 34, 1886.

56. "Lehrbuch der Nervenkrankheiten," 4th ed., Berlin, 1905, II, p. 1313.

tardation and difficulty in active movements and a peculiar modification in gait. The last two are the result of the former, so that really we have two, muscular rigidity and tremor. According to the same author, even the subjective symptoms are due to the muscular rigidity and limitation of active motility. Cases have been reported without tremor, so that it would seem that the muscular rigidity and its peculiar manifestations is the one absolutely characteristic symptom. This rigidity is very different from spasticity, it is not under the influence of the will, is not increased by passive movement, nor is it affected by the mental state of the patient, and it is constantly, gradually progressive, all of which speaks for a muscular affection. Blocq⁴⁶ was probably (so quoted by Redlich²⁴ and by Raymond⁵⁷) the first to formulate the theory of a myopathy as the anatomic basis of the rigidity. He found distinct anomalies in the muscles; irregular atrophy and hypertrophy of the fibers, and proliferation of the sarcolemma nuclei. Joffroy³⁵ had already noted similar changes in the muscles, and Borgherini²⁵ had observed "that in the muscle the connective tissue was increased, the muscle fibers were all somewhat delicate and some were intensely atrophied, showing vacuoles and increase in the number of sarcolemma nuclei." Burzio,²¹ Redlich,²⁴ Pierret and Vessele,⁵⁸ Wollenberg,¹ v. Sass¹⁷ and others found similar changes in the muscles, and by Skoda,⁴ E. Leyden⁵⁹ and Ketscher²⁷ these changes were found combined with a fatty degeneration of the muscles. Dana²⁶ found a fatty degeneration of the muscle which he decided could not be due to neuritis, for the nerve supplying that muscle was normal. In Gordinier's³³ case the muscle was apparently not examined, as he makes no mention of it, and this unfortunately is true in many of the cases reported with necropsy.

Schwenn⁴⁴ described changes in the muscles in his cases similar to those previously described, but called attention to the fact that the changes were not the same in all parts of the muscle, but were at various places at various levels. He comes to the conclusion, on the ground of his own investigation, that the basic anatomic change

57. Article by Raymond: *Nouv. Icon. de l'Salpatrière*, January, 1904.

58. Thèse de Lyon, 1881. Quoted by Raymond, reference 57.

59. *Arch. f. Psych. u. Nervenkrankh.*, vi, 1876, p. 293.

in paralysis agitans is a severe progressive disease of the muscle which gradually involves the whole musculature. He calls attention to the fact that evident changes are found in the muscles during life, and cites Blocq⁴⁶ and Borgherini²⁵ as having found changes in the electric irritability of the muscles. Mendelsohn⁶⁰ observed an increase in the latent period in the diseased muscles. Schwenn⁴⁴ also noted the resemblance to myotonia congenita, "in which many specific changes in the muscles are found," and also that the most common etiologic factors affect the muscles, trauma, strain, excessive use, etc. Shiefferdecker⁶¹ by his very careful pathologic studies, in which comparisons were made with normal control tissue and all the factors of postmortem rigidity, method of hardening, etc., were taken into account, concludes that

Both myotonia congenita and paralysis agitans are diseases of the sarcoplasm and fibrillæ of the muscles. In paralysis agitans, however, there is also a disease of the muscle fibers in the muscle spindles, although the nerve fibers both within and without the muscle spindles are normal.

Idelsohn⁴⁵ describes his findings in the muscles as similar to those of Shiefferdecker,⁶¹ increase in the number of sarcolemma nuclei, hyaline degeneration, swelling of the fibers with vacuolation, etc., and concludes that the anatomic lesion of Parkinson's disease is in the muscles.

I was enabled to examine muscular tissue from 8 of the 14 cases with necropsy; in one other case a piece of muscle was removed from the forearm of a patient in the service of Dr. W. G. Spiller at the Polyclinic Hospital. The patient (E. W.) was 62 years old and had had paralysis agitans about three years. The symptoms of the disease were typical, but otherwise the patient was in good health, and the muscle was removed by me for the purpose of this examination only. I know of but two instances in the literature in which muscular tissue has been removed from the living patient for this purpose: DeBuck and DeMoor,⁶² quoted by Raymond,⁵⁷ and Sanna Salaris,⁶³ "Note di istologia patologica (per biopsia) sulla fibra muscolare striata nel morbo di Parkinson."

60. Arch. de Phys., 1880, p. 218.

61. Deutsch. Ztschr. f. Nervenheilk., 1904, xxv, No. 1-4.

62. Annals de l'Soc. Med. de Gand, June, 1899. Quoted by Raymond, reference 57.

63. Riv. di Patol. nerv. e. Ment., August, 1895, p. 383.

The original of these papers in each instance is inaccessible to me. In all I have examined the muscles from 9 cases of the disease and pathologic changes were present in every case.

A summary of the condition of the muscles is as follows:

Many of the muscle fibers were swollen, and in cross-section were round instead of having the normal polygonal shape. There was a marked increase in the number of nuclei within the fibers, many of which were smaller, denser and rounder than the normal. These nuclei were frequently in pairs or in long chains. In several cases the fibers showed atrophic changes, and in these cases, usually of long-standing disease, there was an overgrowth of the connective tissue. In many cases the fibers had a hyaloid appearance and the longitudinal striations were unusually well marked, and in many of the fibers there was a tendency to longitudinal cleavage. In one case *trichina spiralis* was found in the muscle and in this case, in addition to the above described changes, there was an intense interstitial myositis and also a discoidal degeneration of the muscle fibers. Muscle spindles were hard to find in the sections from any of the cases, but in those examined the muscle fibers within them showed similar changes to those surrounding them. The nerve fiber within the spindle was normal, and the intramuscular nerves showed no degeneration when stained with the Weigert hematoxylin stain. These pathologic changes were not of the same intensity in all the muscles nor in different parts of the same muscle.

My findings in the muscle agree generally with those of Schwenn,⁴⁴ Schiefferdecker,⁶¹ and Idelsohn,⁴⁵ and indicate a definite and probably a specific change in the muscles in paralysis agitans, though the latter can not be regarded as proven. Schiefferdecker⁶¹ regards the lesions as different from those due to overactivity of the muscle. v. Sass¹⁷ examined the muscle of a normal individual 93 years old as a control, and found decided differences between it and the muscle in paralysis agitans. In paralysis agitans the fibers were filled with nuclei, and the small, round, dark-stained nuclei were particularly increased. In the muscle of old age there was an increased number of spindle-shaped and large oval nuclei, but the small dark nuclei were absent.

In a case of paraplegia due to peripheral obliterating arteritis in which I examined the muscles of the legs, I found differences from those in paralysis agitans in the increase of interstitial tissue and in the predominance of the rod-shaped nuclei over the small round nuclei. The fatty degeneration of the muscle fibers found by Skoda,⁴ Dana²⁶ and Ketscher²⁷ was found by the last named also in several patients with arteriosclerosis. I found it present in a case of triplegia following hemiplegia reported by Dr. Charles W. Burr and myself before the American Neurological Association, June, 1906, and in this case it was regarded as due to the lack of nutrition to the muscles from shutting off of the blood supply by an intense peripheral arteriosclerosis. There is not enough known of the pathology of the muscles to say that these changes described are specific in paralysis agitans, but they are co-existent and probably co-extensive with the symptoms rigidity and tremor, though it can not be definitely proven that they are the cause of these symptoms. By some, the changes are regarded as caused by the tremor; the constant contraction of the muscle. But in studying the pathology of disease of other organs of the body we usually attribute the functional changes observed to the lesion that is found if that lesion is adequate to explain the symptoms.

Can we explain the symptoms of the disease by the anatomic findings in the muscles? The onset of the disease is usually gradual, with a gradual progression from muscle to muscle and from part to part, though it follows no regular course, sometimes being hemiplegic, sometimes paraplegic. In this it resembles the rheumatic joint affections more than any other disease, and indeed it has been supposed to be due to similar toxin. Occasionally the onset of the tremor is sudden, but this is explained on the theory of a muscular affection in the same way that a sudden onset is explained by those who hold the opinion that the disease is due to overgrowth of the neuroglia in the cord, etc., and that is that the prodromal symptoms had gone unnoticed. The most characteristic symptom of the disease is the muscular rigidity, and on this symptom most of the other symptoms depend. In the course of the disease it gradually increases and extends in limits so as finally to involve all the muscles in the body except the unstriated. It is not spasticity; the resistance is the same whether the limb be moved quickly

or slowly. It has something of the complete loss of life or elasticity that is felt when a lead pipe is bent. I have found a quantitative change in the irritability of the muscle to the Faradic current in several cases in which the disease was of long duration. "The electric irritability is little affected except toward the end, when there is a quantitative change" (Wollenberg). This, however, is no argument against a local affection of the muscles, as in the muscular dystrophies the electric irritability is equally little affected. The tendon reflexes are usually normal in both diseases, unless the muscles concerned in the reflex become implicated, and the cases in which they are exaggerated in paralysis agitans really prove nothing, since seniles without symptoms of paralysis agitans often show increased knee jerks. All the clinical evidence points to the rigidity as being due wholly to an affection of the muscles. Actual paralysis is very rare in paralysis agitans, and when it occurs is usually limited to one muscle, as in the case reported by Moncorge⁶⁴ of complete paralysis of the right serratus magnus.

Schiefferdecker⁶¹ and others suggest that the pathologic change in the muscle spindles is the anatomic basis of the tremor. It is a very important point that the changes in the muscles in paralysis agitans differ from those found in myotonia congenita only in an involvement of the muscle spindles in the former and not in the latter. Sherrington⁶⁵ says that the muscle spindles are undoubtedly sensory end organs, and Schiefferdecker⁶¹ quotes Baum⁶⁶ as stating that they are organs of muscle sense. Langerhans⁶⁷ comes to the conclusion that they are organs of muscle sense, and that they cause or regulate the muscle tone. As it is generally admitted that the tremor in paralysis agitans is due to a pathologic condition which affects the neuromuscular mechanism that maintains muscle tone, the lesion of the muscle spindles affords a reasonable explanation of the tremor. Another important point tending to prove that there is a relation between the tremor and the lesion of the muscle spindle, and one that I have not seen noted elsewhere, is that the tremor almost always begins in the muscle

64. Lyon Med., No. 6, 1891.

65. Jour. of Physiol., 1894, xvii, p. 237.

66. Anat. Heft, 1900.

67. Virchow's Arch., 1897, cxlix, No. 1.

where the muscle spindles are most abundant, and does not affect those muscles which contain no muscle spindles. Both Millbacher⁶⁸ and Frankel⁶⁹ found the muscle spindles to be most abundant in the muscles that move the thumb. Batten⁷⁰ found them most abundant in the small muscles of the hand and in the biceps. They were not able to find any in the diaphragm or in the masseters muscles. Siemerling,⁷¹ in a case of gumma of the base of the brain, described a rather doubtful muscle spindle in the inferior rectus muscle in one eye, but neither Millbacher, Frankel, Batten nor Sherrington could find any spindles in the ocular muscles, though Sherrington found tendon organs there. William Hirsch,⁷² writing on paralysis agitans in Posey and Spiller's "The Eye and the Nervous System, Their Diagnostic Relations," says:

To the tremor would apparently correspond nystagmus, but this symptom has never been observed in paralysis agitans. This fact furnishes another proof for the assumption that nystagmus is produced by some central lesion and not by a disturbance in the general muscular tonus, because if the latter view were correct we would be bound to observe the symptom at least occasionally in paralysis agitans.

Inferentially, therefore, the tremor in paralysis agitans is due to a disturbance of muscular tonus, which is not operative in the ocular muscles, and the only apparent reason why it should not do so is because of the absence of muscle spindles. M. Koenig,⁷³ Debove⁷⁴ and Galezowski,⁷⁵ writing on the visual or ocular symptoms of paralysis agitans, describe rigidity of the ocular muscles and difficulties in movement due to this rigidity, but do not describe any thing that corresponds to the typical tremor as it occurs in the extremities. Batten⁷⁰ states that he could find no muscle spindles in the intrinsic muscles of the tongue. Forster⁷⁶ found them in the hinder part of the tongue, which muscles, when the two sides act in unison, protrude the tongue and draw it

68. Deutsch. Arch. f. klin. Med., 1882, xxx, p. 304.

69. Virchow's Arch., 1878, lxxiii, p. 380.

70. "Brain," 1897.

71. Arch. f. Psych., 1888, xix, p. 401.

72. "The Eye and the Nervous System; Their Diagnostic Relations." Philadelphia, 1906, p. 477.

73. Bull. Med. 1893, p. 508.

74. Progrés Med., 1878, No. 7.

75. Rec. d. Ophthal., February, 1891, p. 72; also Bull. Med., 1891.

76. Virchow's Archiv., cxxvii, p. 121, 1894.

back. The usual tremor of the tongue in paralysis agitans on protrusion is a piston-like forward and backward movement, and thus here again the peculiar distribution of the muscle spindles bears an apparent relation to the muscles affected by the tremor. The tremor of paralysis agitans has certain peculiarities that are easily explained on the ground that it is due to an affection of the muscle spindles, and in much the same way as they are explained by those who hold that the lesions in the cord are responsible for the tremor. In either case it is admitted that the tremor is due to interference or a change in the impulses that maintain or regulate muscle tone, so that the tremor stops when the muscles are relaxed in sleep; or when a hemiplegia stops them completely on the paralyzed side; or when the impulse for a voluntary act supersedes, momentarily, the regular reflex.

Many of the symptoms of paralysis agitans are easily recognized as due to the muscular tremor and the muscular rigidity, such as the peculiarities in gait, the position, the facies, the ocular symptoms, etc. Other symptoms that are not so obviously due to the two primary symptoms are seen to be so on a careful analysis, thus Catola⁷⁷ comes to the conclusion that the salivation is due first, to tremor of the lips, cheeks and tongue; second, to rigidity of the same organs and of the muscles of the pharynx, with more or less complete interference with the movements of deglutition. The recumbent position of the patient permits the saliva to descend to the posterior part of the pharynx and so stimulates the reflex for deglutition. Other conditions often arise in paralysis agitans which are by some regarded as symptoms, but which might more reasonably be looked on as complications. M. A. Soques⁷⁸ has recently called attention to the syndrome of labioglossolaryngeal palsy occurring as a symptom of paralysis agitans, and quotes Bruns⁷⁸ as having found it in 4 cases in 74. There are not the characteristics of a true bulbar palsy, but rather a pseudo-bulbar palsy. Dejerine, in a discussion of this paper, considers the syndrome as a complication and called attention to the fact that many patients with paralysis agitans also have arteriosclerosis and are in a condition favorable for the production of a pseudo-bul-

77. *Rev. Neur.*, 1905, xlii, p. 138.

78. Article by Soques, M. A.: *Rev. Neurol.*, 1905, xlii, p. 135.

bar palsy. The parasthesiæ, the sensations of heat and cold might well be considered as complications due to the age of the patient rather than symptoms of the disease. In an analysis of 219 cases, Hart⁷⁹ found sensations of heat in only 31, sensations of cold in 20, and both kinds in 20. Sensations of heat and cold are not at all uncommon in many people of advanced age.

It may be that the same factor which produced the changes in the muscles also makes itself felt in other ways, in producing a thickening or a loss of elasticity in the skin, for instance. Frankel,⁸⁰ in calling attention to the loss of elasticity of the skin in paralysis agitans, says "that perhaps the muscle fibers are altered by the same influence." A. M. Luzzato⁸¹ found that in a case of paralysis agitans combined with a scleroderma, both were improved for a time by the administration of arsenic. A histologic examination threw no light on the pathogenesis of the condition, but he concludes that it was a double expression of one morbid state.

THE PATHOGENESIS OF THE DISEASE.

Two general hypotheses may be considered as to the underlying cause of the changes in the muscles: First, there may be a primary degeneration of the muscles themselves, an abiotrophy, affecting the muscles analogous to that in the muscular dystrophies. Beginning in a muscle frequently after some slight local exciting cause, it gradually progresses from part to part. It may well be that people are born with muscles which lose their elasticity more quickly than the muscle of other people, either by use or through other causes that we know little about. The property of elasticity of a muscle has been shown experimentally to be lost by over-extension and is subject to various influences.

A second hypothesis is that there is a toxin circulating in the body which causes these changes in the muscles. The idea of a toxin as the underlying pathogeny of the disease has been widely held. Dana²⁶ says:

Perhaps the primary infection in paralysis agitans is some autochthonous poison allied to that of rheumatism; at any rate there is no other preceding history that is so often obtain-

79. Jour. of Nerv. and Ment. Dis., 1904, v, No. 31, p. 177.

80. Deutsch. Ztschr. f. Nervenheilk., 1899, p. 430.

81. Il Morgagni, 1904, p. 753, abs. in Rev. Neurol., 1905.

able as that of rheumatism, and there are many facts in the clinical history of the disease which suggest a relationship between paralysis agitans and that form of rheumatic manifestation which is known as rheumatoid arthritis.

Spiller⁸² has called attention to the points of resemblance between paralysis agitans and rheumatoid arthritis.

The examination of the excretion in paralysis agitans has produced some results of interest. Gauthier⁸³ found an increase of phosphoric acid in the urine in cases of paralysis agitans which suggested to him that the muscular changes may be due to an excess of the same in the muscles. His conclusions were that

The symptoms are due exclusively to the changes in the muscular system and derived from the muscular rigidity. This rigidity, the most important phenomenon of the disease, is produced in the muscles without the intervention of the nervous system and is due to an excess of phosphates.

Cheron⁸⁴ had called attention in 1877 to the increased amount of phosphates in the urine, but by him they were considered as affecting the central nervous system, and so producing the symptoms of the disease. G. Gurtler,⁸⁵ C. A. Ewald,⁸⁶ Giovanni Setti⁸⁷ and Decombrel⁸⁸ found no increase of phosphates in the urine. Researches in the contents of the urinary excretion in paralysis agitans have also been carried out by Mosse and Baul,⁸⁹ who found an increase of the completely oxidized phosphates, but a corresponding diminution in the incompletely oxidized; and by J. Leva⁹⁰ and R. Bandeio,⁹⁰ who found it negative.

In poisoning by phosphorus (Siemerling,⁹¹ also Langerhans⁹²), the muscle fibers within the muscle spindles are affected, as well as those outside, which is similar to the effect produced in paralysis agitans. It may be well here to call attention to the remarkable immunity enjoyed by the muscle spindle in various

82. Univ. Penn. Med. Bull., May, 1904.

83. Lyon Med., 1888, No. 35-36.

84. Le Progrès Med., 1877, No. 48.

85. Arch. f. Psych., xiv, 1888.

86. Berlin. klin. Wochschr., 1883, No. 32-33.

87. Arch. Ital. di clin. Med., xxxvi, p. 262. Quoted by Schwenn, reference 44.

88. Rev. de Med., July, 1889, p. 583.

89. Deutsch. Ztschr. f. Nervenheilk., 1892, II, No. 1, p. 75.

90. Gaz. hebdom. de Sci. Med., de Bord., 1905, xxvi, 291-293.

91. "Charité Annalen," 1889, p. 443.

nervous affections, even when the muscle itself is severely atrophied or even in primary muscle affections, such as the muscular dystrophies. Roth⁹² found them normal in a case of alcoholism, a case of progressive muscular atrophy, and in a woman who died of inanition consequent on a pyloric stenosis. Siemerling,⁹³ in a case of spinal neuritic muscular atrophy, in which he found an intense atrophy of the muscles and a complete degeneration of the intramuscular nerves, found the muscle spindle "entirely unchanged." Blocq and Marinesco⁹⁴ found them present in cases of poliomyelitis and polyneuritis when there was total degeneration of all the other muscle fibers. Spiller⁹⁵ found them normal in a case of of intense progressive muscular dystrophy with contractures, in which there was no other normal muscular tissue. Grünbaum,⁹⁶ in a case of pseudo-muscular hypertrophy, with great changes in the muscles, found that both muscles fibers and the nerve within the spindles were normal. V. Horsley⁹⁶ found the muscle spindles merely shrunken after division of the nerve in dogs. This immunity of the muscle spindle in degeneration of the muscle consequent on nervous lesions and in the ordinary muscular dystrophies is in marked contrast to the decided effect produced by toxic substances, but there is not sufficient evidence, and what there is is too contradictory to uphold the opinion of Gauthier that the degeneration of the muscles and muscle spindles in paralysis agitans is due to phosphoric acid or one of its compounds. The idea that it is due to some toxic substance, however, receives confirmation.

Under the head of circulating toxic substances may be classed defective or changed internal secretions. Fibrous changes in the ovaries or testicles are regularly found at necropsy in cases of paralysis agitans, but they show no differences from the changes of senility, which in all probability they are. The extirpation of the testicles or ovaries produces no symptoms in any way similar to paralysis agitans, and the injection of such substances as Brown-Séquard extract, on purely empirical grounds it would seem is, so far as I know, of no benefit in this dis-

92. *Centralbl. f. die med. Wissensch.*, 1887, p. 129.

93. *Arch. f. Psych. u. Nervenkrankh.*, **xxi**, No. 1-2, p. 105.

94. *Soc. de Biol.*, 1890, p. 398. Quoted by Batten, reference 70.

95. *Jour. Nerv. and Ment. Dis.*, October, 1897.

96. *Brain*, Autumn, 1897.

case. Moebius,⁹⁷ in 1883 (quoted by Raymond⁵⁷) reported a case of paralysis agitans combined with Basedow's disease and opined that paralysis agitans was due to an alteration in some glandular secretion. Lundborg⁹⁸ reported a case of paralysis agitans combined with myxedematous symptoms, and at necropsy found atrophy and cystic changes in the thyroid, which led him to believe that the thyroid gland plays an important part in paralysis agitans. Langerhans⁶⁷ found that in cretins there was a fatty degeneration of all the muscles except the tongue, cricothyroid, thyrohyoid, and rectus abdominus muscles which was similar to the general fatty degeneration of the muscles seen in phosphorus poisoning and in which the muscles fibers in the muscles spindles also took part. Askanazy,⁹⁹ in a pathologic study of Basedow's disease with special reference to the muscle changes in the disease, found that the muscles fibers were atrophied and had undergone a fatty metamorphosis, the striations were lost and the fibers appeared homogeneous, there was vacuolation of the muscle fibers and proliferation and degenerative changes in the sarcolemma nuclei. The muscles fibers in the muscle spindles showed the same changes as those about them. He noted a marked tremor of the hands in his case. Castelv¹⁰⁰ reported pathologic alterations of the thyroid in two cases of paralysis agitans. There are several facts against any change in the internal secretion of the thyroid gland operating as the cause of paralysis agitans. The symptoms which accompany marked disease or even total destruction of the gland are not similar to those of paralysis agitans; Alquier¹⁰¹ has examined the thyroid and adrenal glands from cases of paralysis agitans and found not the slightest alteration. In the two cases in which I had examined the thyroid glands there was a simple colloid degeneration in each case, not accompanied by gross enlargement of the gland, and probably a senile change. Thyroid therapy has generally been barren of results.

The numerous cases reported of a combination of

97. *Memorabilien*, 1883. Quoted by Raymond, reference 57.

98. *Deutsch. Ztschr. f. Nervenheilk.*, 1901, xix, p. 268.

99. *Deutsch. Arch. f. klin. Med.*, 1898, lxi, p. 118.

100. *Riv. de Med. y P Cirugia practicas*, Madrid, 1903; abs. in *Rev. Neurol.*, 1904, p. 141.

101. *Thèse de Paris*, 1903; *Rev. Neurol.*, 1904, p. 438.

paralysis agitans with symptoms of myxedema or Graves' disease and the fact that the muscle changes observed by Askanazy⁹⁹ are similar to those in paralysis agitans lead to the conclusion that there is some intimate relation in their causes and as a logical result to an investigation of the parathyroid glands. The parathyroid glands have gradually assumed a position of great importance in pathology since their discovery by Sandstrom. They were at first supposed to be accessory or embryonic thyroids. Welch (cited by Richardson¹⁰²) considers that anatomically the parathyroid glands resemble the anterior lobe of the pituitary body, and that unlike the thyroid the presence of a colloid material in the gland is retrograde or degenerative in its nature. Richardson¹⁰² quotes Gley, an early investigator of the subject, as holding the opinion, supported by physiologic experiments, that the parathyroids are glands *sui generis* and have a specific function related to that of the thyroids. That the removal of the thyroids in one case and the parathyroids in another produces very different results is also proven. In a personal communication Dr. W. S. Carter, now of the University of Texas, Galveston, from his experiments performed on dogs while at the University of Pennsylvania, concludes that "the parathyroids are more important than the thyroids, that the acute nervous symptoms in dogs which follow total extirpation of both thyroids and all four parathyroids are due to the removal of the latter bodies and not to the former. Total parathyroidectomy without thyroidectomy produces tetany and death just as certainly as the total extirpation of both sets of bodies. On the other hand, extirpation of the two thyroids, leaving one or two external parathyroids with the circulation intact may cause transitory symptoms from the accidental injury to the parathyroids at the time of operation, but I have had such animals live for months in perfect health. The subsequent removal of the remaining one or two external parathyroids would then produce the characteristic tetany and death within a few days." These results are in accord with those obtained by other investigators.

Richardson¹⁰² gives the acute symptoms in a parathyroidectomized dog as tremor, rigidity, convulsions,

102. "The Thyroid and Parathyroid Glands," Philadelphia, 1905.

dyspnea, and death. With due allowance for the sudden onset and complete removal of the gland in these experiments compared to the more gradual destruction of the gland by a pathologic process, from all the descriptions given, the symptoms presented by a parathyroidectomized animal are very similar to those of paralysis agitans. In the picture of a monkey in this tetanic condition (v. Eiselberg reproduced by Richardson¹⁰²) there are seen many resemblances to the characteristic position of a patient with paralysis agitans: the head and back bent forward, the joints of the extremities in flexion, the general appearance of rigidity and even the ulnar deviation of the hands. So far as I know, Lundborg¹⁰³ in 1904 was the first to call attention to the resemblance between the symptoms of paralysis agitans and the effects of parathyroidectomy in animals, and he arrived at the conclusions that "paralysis agitans is probably a chronic, progressive hypoparathyroidismus." Berkley¹⁰⁴ also noted this similarity in symptoms and reports eleven cases of paralysis agitans treated with parathyroid extract, nine were benefited and he refers to one very early patient who considered himself cured. These results of Berkley assume more importance in throwing light on the pathogenesis of the disease when we recollect the uniformly discouraging results of treatment with the other glandular extracts. Neither Lundborg or Berkley had any pathologic evidence to support their conclusions.

I was able to examine the parathyroid glands from two of my cases, and in the opinion of Dr. Allen J. Smith, professor of pathology at the University of Pennsylvania, they were both in a distinctly pathologic condition. In one there was some colloid material which, according to Welch, indicates a degenerative condition, but in both these cases there was a peculiar infiltration with fat, especially in relation to the blood vessels. This pathologic evidence of disease of the parathyroid in paralysis agitans, when considered together with the experimental evidence quoted and the therapeutic results of Berkley, suggests that the parathyroid glands play an important part in the pathogenesis of paralysis agitans.

103. *Deutsch. Ztschr. f. Nervenheilk.*, 1904.

104. *Med. News*, Dec. 2, 1905.

CONCLUSIONS.

1. Paralysis agitans is not a neurosis nor is it senility.

2. The anatomic basis of the symptoms, muscular rigidity, tremor, and the symptoms dependent on them, lies in the affection of the muscles.

3. The disease itself is probably a general toxemia and there is suggestive evidence that it is due to alteration in the secretion of the parathyroid glands.

I have to thank Drs. W. G. Spiller, Charles K. Mills, Charles W. Burr, James Hendrie Lloyd, Charles S. Potts, D. J. McCarthy, T. H. Weisenburg, and J. H. W. Rhein for referring to me the pathologic material on which this research is based, and especially to thank Dr. William G. Spiller and Dr. Allen J. Smith for verifying the pathologic findings.

CASE REPORTS.

CASE 1.—Patient, J. R., male, aged 69, was admitted to the Philadelphia Hospital, Dec. 22, 1894; died April 7, 1906.

Diagnosis.—Paralysis agitans. Case of Dr. J. H. Lloyd.

Clinical History.—Patient was a laborer. His grandfather, grandmother and aunt had a similar tremor. He had variola at 16 years of age and syphilis at 40 years of age, but no other illness and there was no history of a traumatism. He had used tobacco and alcohol to excess. He said that the tremor began four years ago; first in the head, then in the arms and then in the legs. In 1902, it was noted that he walked with his body bent forward but with no festination. There was a constant tremor of his lower jaw and a coarse tremor of his arms and legs. The tongue on protrusion showed a piston-like movement. The tremor of the hands was of the pill-rolling type and was made worse after voluntary movement. It was more marked on the left side. The facial expression was wooden and there was rigidity of the arms and legs. There was no parathesiæ. Knee-jerks were much diminished.

Jan. 23, 1906: His condition was much the same and he at no time improved. His hands were usually held in the obstetric position. His knee-jerks were lost, and he had incontinence of urine and feces. There was no disturbance of articulation nor any nystagmus.

Feb. 6, 1906: He developed delusions of persecution, was disorientated and confused. He complained of constant pain in his head. The tremor ceased when he slept.

April 5, 1906: The patient's articulation was very poor but his mental condition seemed about normal. The tremor was of the kind characteristic of paralysis agitans; was slightly more

pronounced in the left hand and stopped at times when the patient was very quiet. There was a marked atrophy of the interossei muscles, particularly the first, in both hands, but more marked in the left. There was an ulnar deviation of both hands but the skin of the hands was normal and the small joints were not enlarged.

Death was sudden and due to uremia. At autopsy a chronic interstitial nephritis and general arteriosclerosis was found.

Pathologic Examination.—Macroscopically, the appearance of the general nervous system was normal.

Microscopically, sections of the paracentral lobule appeared normal. The Betz cells were much pigmented. The medulla and cerebellum appeared normal when stained with the hemalum acid fuchsin, the Weigert hematoxylin or the Mallory neuroglia stains. Sections of the cervical cord showed increased prominence of the capillaries. The spinal canal was closed and there was a large number of amyloid bodies in the spinal cord, especially in the posterior columns. Sections from the thoracic and lumbar regions showed the same condition as those from the cervical. The increased prominence of the capillaries was most marked in the thoracic region. The anterior horn cells, stained by the thionin method, were much pigmented, and a few showed distinct chromatolytic changes. Sections of both median and both posterior tibial nerves appeared normal when stained with the hemalum-acid fuchsin and Weigert hematoxylin stains. No degeneration could be detected with the fresh osmic acid stain nor with the Marchi method.

In cross sections of the right biceps muscle the fibers appeared swollen and round. There were no vacuoles in the fibers. In longitudinal sections an increased number of sarcolemma nuclei was noted. There was also, within the fibers, a large number of nuclei, probably also sarcolemma nuclei, but smaller and denser and round instead of the normal oval. They were frequently in pairs but sometimes in long chains. The transverse striations were generally indistinct and in some fibers were lost. There did not seem to be any increase of the fibrous connective tissue between the fibers. The intramuscular nerve fibers stained well with Weigert hematoxylin stain. Sections of the left biceps, the right and left adductors pollicis, and the right and left posterior tibial muscles, showed the same changes with but slight variations in intensity. Sections of the adrenal glands showed them to be normal. The thyroid gland contained an excessive amount of colloid material and the epithelial lining of the acini was much thinned. Both parathyroid glands were the seat of well-marked injection of the vessels and there was also an increased thickness of some of the vessel walls. There were a number of microscopic foci, in the left particularly, in which the parenchyma was absent and replaced by fat and this was particularly to be noted along the course of the thickened vessels. It was the opinion of Dr.

Allen J. Smith, professor of pathology at the University of Pennsylvania, that the changes in the parathyroid glands were distinctly pathologic.

CASE 2.—M. McL., female, aged 53, admitted to the Philadelphia Hospital April 23, 1897; died, Jan. 19, 1905.

Diagnosis.—Paralysis agitans. Case of Dr. Charles W. Burr.

Clinical History.—Laundress. Present trouble began five years previous to her admission to the hospital, with pain in the left shoulder and tremor of the left hand. This stopped in two or three years and she returned to her work. About one year before her admission the tremor began again in both hands, she attributed it to her work, and at the time of her admission was rhythmic in her head and in both hands. Flashes of heat through her body began soon after the tremor and have persisted ever since. She sat with her head and back bent forward and her face was expressionless. There was distinct tremor of the tongue but none of the lips. Knee-jerks were diminished.

Oct. 30, 1903: There was deformity of the left hand, i. e., the thumb flexed; the hand flexed at the metacarpal joints; the proximal phalanges extended, and the terminal were flexed, but could be extended by an effort. Sensation remained normal throughout. Death was due to uremia.

Pathologic Examination.—Macroscopically, the appearance of the central nervous system was normal.

Microscopically, sections from the cervical regions of the spinal cord showed an apparent increase in the number of blood vessels, especially in the posterior columns, due to a thickening of their walls. There are no distinct islets of glia about the blood vessels. Amyloid bodies were numerous about the periphery of the cord. The perivascular spaces are considerably widened, especially in the gray matter. The Mallory neuroglia stain showed a moderate subpial thickening and also an overgrowth of glia in the cord, but not excessive. Sections from the thoracic and lumbar regions present a very similar appearance. There was no degeneration in the spinal cord detectable by the Weigert hematoxylin or the Marchi methods of staining. The anterior horn cells of the spinal cord appeared generally normal when stained by the thionin stain. They contained a large amount of pigment and a few showed chromatolytic change. The anterior and posterior nerve roots were congested but appeared otherwise normal when stained by the acid fuchsin and Weigert stains. Sections from the paracentral lobule showed some of the Betz cells with chromatolytic changes but most of them were normal and not excessively pigmented. The sections were otherwise normal. Sections from the medulla, pons and cerebellum showed them to be normal. The twelfth thoracic posterior root ganglion was found to be normal except for an increased amount of connective tissue.

The nerve cells were normal. The obturator nerve was found to be normal except for a moderate increase in the connective tissue between the nerve bundles. In sections of the anterior crural nerve the medullary sheaths appeared swollen and granular. The posterior tibial muscle, cut transversely, was seen to be distinctly atrophied; though not equally so in all parts of a section. The amount of fibrous tissue was increased. In one part of a section the fibers were small, rounded off and widely separated, one from another, in a different part the fibers were even larger than normal, were ill defined, and contained vacuoles of various shapes and sizes. No muscle spindles were observed in the sections. An intramuscular nerve filament showed no degeneration when stained by the Weigert method. The anterior tibial muscle may be described as in the same condition as the posterior tibial though there was a great predominance of the smaller fibers seen in the sections. One muscle spindle observed showed three fibers cut transversely that were distinctly striated and apparently normal; the muscle fibers in that part of the section immediately surrounding the spindle appeared normal. In another muscle spindle in the section the muscle fibers appeared distinctly atrophied. The intramuscular nerves stained normally with the Weigert hematoxylin stain. Changes in the peroneal muscle were of the same character as already described in the tibial; some of the fibers cut longitudinally, showed a longitudinal cleavage. Sections from the intercostal muscles and the diaphragm showed much the same changes but to a less extent. The pancreas were normal. The adrenals were normal except for a slight cloudy swelling of the parenchymal cells. Sections of the ovaries showed them to be completely cirrhotic, not a trace of the normal structure remaining. The walls of the blood vessels were intensely sclerotic with the lumen almost obliterated. The thyroid gland was not grossly enlarged but sections showed an excessive amount of colloid material and a distinct thinning of the epithelial layer lining the acini. The parathyroid glands showed considerable congestion and sclerosis of the blood vessels. There was a number of microscopic foci in which the parenchyma had disappeared and was replaced by fat, particularly along the blood vessels. In some of the cells the protoplasm appears swollen and does not stain deeply, probably a mucoid change. In the opinion of Dr. Allen J. Smith, professor of pathology at the University of Pennsylvania, the changes in the gland were distinctly pathologic.

CASE 3.—E. M., female, white, aged 65, was admitted to Philadelphia Hospital Nov. 29, 1900; died June 30, 1905. Patient did housework; was married, but never pregnant.

Diagnosis.—Paralysis agitans. Case of Dr. D. J. McCarthy.

Clinical History.—On June 2, 1904, it was noted: That she began to have "rheumatism" ten years ago. The tremor first appeared in the left hand six years ago, several months later

in the left leg, a year later in the right arm, and last in the right leg. There is a marked tremor of both hands with the characteristic pill-rolling movement which is decreased by voluntary motion. She stands with her body bent at almost a right angle and can walk slowly unsupported with eyes open or closed. There is a constant flexion and extension of the right foot. There is rigidity in all of the limbs. She has very little power in the legs, can only raise them a short distance from the floor and is weaker in the right arm than in the left. She has had at times a burning sensation in the legs, back and head. Her tendon reflexes are prompt and equal. There is no Babinski reflex and sensation is normal. There are periods of absence of winking followed by quickly repeated winks, but no other ocular symptoms. Death was due to nephritis.

Pathologic Examination.—Macroscopically, the appearance of the central nerve system was normal.

Microscopically, sections from the paracentral lobules appeared considerably congested but otherwise normal. The tangential fibers stained well by the Weigert hematoxylin stain. The Mallory neuroglia stain showed nothing abnormal. The Betz cells, stained with thionin, appeared normal, a few showed excessive pigmentation. Medulla, pons and cerebellum were normal. Sections from the cervical region of the spinal cord showed numerous amyloid bodies, especially in the posterior columns. The capillaries appeared very numerous and prominent, probably because of the thickening of their walls: the Mallory neuroglia stain showed no overgrowth of glia about them. Sections from the thoracic and lumbar regions had much the same appearance as those from the cervical. There was no degeneration in the spinal cord detected by the Weigert or by the Marchi methods. The anterior horn cells were intensely pigmented and the chromaphilic substance stained somewhat diffusely by the thionin stain but the nuclei and the cell processes were normal.

Sections from a lumbar posterior root ganglion showed an overgrowth of connective tissue but no degeneration of the nerve fibers. The cells contained large quantities of coarsely granular pigment, and are rounded, appearing swollen, with a finely granular chromaphilic substance.

In sections of muscle from the leg there was seen a marked increase in the number of sarcolemma nuclei which were arranged in long chains. There was little, if any increase, in the amount of fibrous tissue. The transverse striations of the muscle fibers were generally well preserved and the longitudinal striations were very prominent. Some of the fibers appeared much convoluted on longitudinal section and showed a tendency to longitudinal cleavage. A granular pigmentation is seen in many of the fibers. A muscle spindle cut transversely, showed atrophy of the muscle fibers within it. The intramuscular

nerve fibers stained well by the Weigert hematoxylin method, as did also the nerve fibers within the muscle spindle.

CASE 4.—J. U., male, aged 54.

Diagnosis.—Paralysis agitans. Case of Dr. J. H. W. Rhein.

Clinical History.—At an examination by myself made Jan. 30, 1903, there was noted the following: He was an insurance agent. The family history is negative, except that a brother committed suicide two years before. There is no history of venereal disease or of alcoholic excesses. For the past 30 years he has suffered from severe migrainous attacks which would come on just before a storm. He had an infected arm 30 years ago. The tremor began suddenly, 14 months ago, immediately after a severe fright. It first affected the left arm and two months later the left leg. He felt cold very acutely but had no pain or other symptoms. On examination it was seen that he stood with his knees and back bent. His face was immobile, but there was a tremor of his head. The left arm and leg were both rigid, the arm more than the leg, but there were no fibrillary twitchings in the muscles and the electrical reactions were normal. The tendon reflexes of both upper and both lower extremities were exaggerated. There was no Babinski reflex nor ankle clonus and sensation was normal. The tremor was momentarily stopped by voluntary movements. Before his death the tremor had affected the right arm and leg also, and he complained a great deal of pain in the muscles of the arms and legs. Death was due to uremia and occurred in February, 1906.

Pathologic Examination.—Macroscopically, the appearance of the central nervous system was normal.

Microscopically, sections from the paracentral lobules appeared normal by the hemalum acid fuchsin and Weigert hematoxylin stain. The Betz cells appeared normal by the thionin stain. The pia of the spinal cord was slightly thickened and the anterior spinal artery much sclerosed. The intramedullary capillaries were very prominent because of the thickening of their walls. There was slight subpial glial hypertrophy. The spinal canal was open. There was no degeneration detected by the Weigert method nor by the Marchi method for recent degeneration.

Sections from the thoracic and lumbar regions showed much the same changes. The anterior horn cells generally appeared normal when stained by thionin; a few showed degenerative changes and many contained large amounts of pigment. Sections of nerve from the leg showed it to be normal. Sections of a muscle from the leg showed numerous trichina spiralis in a late stage of their development. The muscle fibers were exceedingly wavy, and irregularly contoured. Many of the fibers had split into discs within the sarcolemma sheath which continued unbroken. These discs take the Weigert hematoxylin stain very intensely. Many of the fibers had a hyaloid appearance and

the transverse striations were indistinct. The sarcolemma nuclei were increased in number but there was also large numbers of smaller, round nuclei within the fibers. Many of the fibers contained vacuoles. There was a considerable increase in the amount of connective tissue, especially about the blood vessels, which was rich in nuclei (interstitial myositis). No muscle spindles were seen in the sections. The intramuscular nerves stained well by the Weigert hematoxylin method.

CASE 5.—Age unknown.

Diagnosis.—Paralysis agitans. Case of Dr. T. H. Weisenburg. Clinical history not obtained.

Pathologic Examination.—Macroscopically, the appearance of the central nervous system was normal.

Microscopically, sections from the paracentral lobule were normal. Sections from the cervical region of the spinal cord showed an increased amount of glia in both columns of Goll and in both lateral columns, especially near the gray matter. It had a diffuse arrangement but was not confined to any system of fibers nor was it in any way connected with the blood vessels. The blood vessels were not sclerotic and there was no evidence of inflammation. The anterior horns were congested. The central canal was open and there was no proliferation of the ependyma. The anterior horn cells, stained with thionin, appeared normal. An overgrowth of the glia, similar to that in the cervical region, was also seen in the thoracic and lumbar regions in sections stained with the hemalum acid fuchsin stain and by the Mallory neuroglia stain. There was no degeneration in the spinal cord detected by the Weigert or Marchi methods. The anterior and posterior nerve roots were normal.

Sections of the median nerve stained by the hemalum-acid fuchsin method showed a marked swelling of the myelin sheaths of some of the fibers. In many of the fibers the axis cylinders had disappeared; in others they are much swollen. The connective tissue between the nerve bundles was moderately increased. The nerves did not appear degenerated when stained by the Weigert method. Sections of muscle showed some fibers that appeared swollen; many were normal but often lying between normal fibers; there were thick ones in which the longitudinal striations were very prominent. There was a marked increase in the number of sarcolemma nuclei; often they lay in long chains of six or more. The intramuscular nerves stained normally with the Weigert hematoxylin stain.

CASE 6.—I. B., male, aged 68, blacksmith, was admitted to the Philadelphia Hospital Dec. 4, 1901; died Jan. 21, 1902.

Diagnosis.—Paralysis agitans. Case of Dr. Charles K. Mills.

Clinical History.—Tremor of both hands began four years previous to his admission to the hospital. It ceased when he made voluntary movements. There was a coarse tremor of the tongue. His speech was slow and drawling and his face expres-

sionless. He walked bent forward with his knees flexed. Kneejerks were "practically" absent.

Pathologic Examination.—Macroscopically, the appearance of the central nervous system was normal.

Microscopically, sections from the cervical region of the spinal cord showed numerous minute hemorrhages into the gray substance and to a less extent into the white matter. They were of recent date as the corpuscles were not altered. Similar, but much less distinct hemorrhages, were seen in sections from the lumbar region. The walls of the small blood vessels were much thickened at all levels of the cord. There were no signs of a meningitis about the spinal cord but there was a slight, round-cell infiltration around the foot of the cerebral peduncles. The spinal roots were normal. There were no signs of any hypergliosis in the spinal cord nor was any degeneration detected by the Weigert hematoxylin method. The cells of Betz in the paracentral lobule and the anterior horn cells of the spinal cord appeared normal when stained by the thionin method, although considerably pigmented.

CASE 7.—A. M., female, aged 45, was admitted to the Philadelphia Hospital Oct. 10, 1898; died Dec. 11, 1904.

Diagnosis.—Paralysis agitans with epilepsy. Case of Dr. C. K. Mills.

Clinical History.—Fifteen years before her admission to the hospital she fell unconscious on the street. Again, three years ago, she had a similar attack, and when she recovered consciousness she had a "weakness" of the left arm and leg and a tremor of the left arm, which has since increased. One year later she developed a tremor of the left leg and a festinated gait. Epileptic attacks began April 5, 1899 and were diagnosed by Dr. C. W. Burr as "petit mal."

July 2, 1899: Severe pain in the back was suddenly followed by a paraplegia with relaxation of the sphincters. Temperature, 104 $\frac{3}{5}$, pulse 106, respiration 34. Next day she could move her limbs but had no control of her sphincters; was up and about in two weeks.

March 3, 1901: Patient had deformed knees with great bony enlargement of the tibia and had severe pains in the legs and feet. Face was expressionless.

June 24, 1901: Eye examination made by Dr. Hansell showed: "central retino-choroiditis of the right side; incipient atrophy of both nerves, but no choroiditis of the left."

Jan. 5, 1903: Tremor persisted during the epileptic seizures, which were becoming more frequent.

Dec. 10, 1904: She had an attack of pneumonia at which tremor stopped.

Pathologic Examination.—Macroscopically, the appearance of the central nervous system was normal.

Microscopically, sections from the paracentral lobules ap-

peared much congested. The Betz cells were greatly pigmented and some appeared to be diseased. They were smaller than their usual size; the nuclei in many were peripherally situated, and the chromaphilic elements were more or less disintegrated. The endothelial cells lining the dura on the dural aspect were proliferated in places. Medulla and cerebellum were normal. Sections of the spinal cord from the cervical, thoracic and lumbar regions showed an immense number of amyloid bodies, especially in the posterior columns. The pia was thickened and showed a moderate round-cell infiltration. The blood vessel walls were thickened but there was no overgrowth of the glia about them. No degeneration in the spinal cord could be detected by the Weigert method. The anterior horn cells appeared normal, though already pigmented, when stained by the thionin method. Anterior and posterior spinal nerve roots, seen in the section, appeared normal.

CASE 8.—C. McM., female, aged 75, was admitted to the Philadelphia Hospital Aug. 7, 1899; died Dec. 2, 1899.

Diagnosis.—Paralysis agitans. Case of Dr. C. K. Mills.

Clinical History.—Rigidity of left arm, came on slowly, beginning two years before her admission; face was mask-like; ocular movements normal; had double senile cataract; had festinated gait; evidence of a fracture of the left forearm; skin of the legs was rough and coarse and there was considerable rigidity in them; her movements were slow and wax-like; there was a fine tremor of the lips and at times of the head; a coarse tremor of the hands and occasionally of the left leg which was worse on excitement and ceased on willed movements; speech was slow, becoming faster after talking a while; tendon reflexes were equal and normal on each side; sensation normal.

Dec. 2, 1899: Condition was the same.

Pathologic Examination.—Macroscopically, the appearance of the central nervous system was normal.

Microscopic sections of the cervical region of the spinal cord showed conspicuous and much distended blood vessels, also some recent hemorrhages in the anterior horns which were probably agonal. There was marked arteriosclerosis of the cerebral and spinal vessels and they were intensely congested, but there was no gliosis about them. No changes were detectable by the Weigert hematoxylin method in the spinal cord and there were no spaces from which fibres have disappeared. Their spinal nerve roots were normal. The medulla, the pons and the cerebellum appeared normal by the hemalum-acid fuchsin and Weigert hematoxylin stains. The Betz cells of the paracentral lobules were much pigmented and the nerve cells of the anterior horns of the spinal cord were moderately pigmented but appeared otherwise normal. The posterior tibial nerve from the left side appeared moderately, but distinctly degenerated, when stained by the Weigert hematoxylin method. Sections from the right tibialis

posticus muscle show it to be about normal. Sections from the left posterior muscle (tibial) showed distinct interstitial myositis; increase of connective tissue, fibrosis about the blood vessels, and increase of the connective tissue nuclei. In some of the fibers there was a distinct increase in the number of the sarcolemma nuclei which were formed in chains. A few vacuolated fibers were observed, but none with a hyaline appearance. The transverse striations were generally well preserved. In some of the fibers the longitudinal striations were very marked. Similar, but less marked changes, were noted in the left biceps muscle.

CASE 9.—J. O'N., male, aged 52, was admitted to the Philadelphia Hospital April 14, 1906; died April 27, 1906.

Diagnosis.—Paralysis agitans. Case of Dr. C. H. Potts.

Clinical History.—He had gonorrhea three times and syphilis, 25 years before his admission to the hospital; otherwise, he had always been healthy. He stated that he never had had rheumatism. The tremor began in his left thumb five years previous to admission. It then affected the rest of that hand; then the right hand; then the right leg, and, finally, one year ago, the face. On examination there was found a coarse tremor of both thumbs, particularly the left, consisting of adduction and abduction, with a slight flexion and extension movement of the first fingers, though they were kept extended. The tremor ceased on voluntary motion or when the arm was supported. The tendon reflexes were normal and there was no ankle clonus nor Babinski reflex. The face was immobile and there was a tendency to festination in his gait. There was distinct rigidity of both arms and slight rigidity in both legs. Death was due to acute dilatation of the heart.

Pathologic Examination.—Macroscopically, the appearance of the central nervous system was normal.

Microscopically, sections from the paracentral lobules appeared normal. The Betz cells, stained by the thionin method appeared normal, though they were considerably pigmented. Sections of the cerebellum and medulla were normal. Sections from the spinal cord showed no sclerosis, either in the larger blood vessels or of the capillaries. There was no overgrowth of neuroglia in the spinal cord nor was there any degeneration detected by the Weigert method. The anterior horn cells, stained by the thionin stain appeared normal and were slightly pigmented. The central canal of the spinal cord was closed by proliferated ependyma. There were no amyloid bodies in the spinal cord.

CASE 10.—T. W., male, aged 65, was admitted to the Philadelphia Hospital June 23, 1905; died July 17, 1905.

Diagnosis.—Paralysis agitans. Case of Dr. C. H. Potts.

Clinical History.—Tremor began five years before admission to hospital. Face was expressionless. There was festination of gait. The hands were held in the obstetric position and

there was marked muscular rigidity. The tremor was not distinct but at times, especially after voluntary movements of the hand, there were five or six coarse, to-and-fro movements of the thumb. Knee-jerks were prompt.

Pathologic Examination.—Macroscopically, the appearance of the central nervous system was normal.

Microscopically, sections from the cervical region showed an apparent increase in the number of capillaries, probably because the walls of the finer ones were thickened. Most of the nerve cells were normal, some showing a slight chromatolysis. The thickened blood vessels were still more distinct in the thoracic region and similar changes were noted in the lumbar region. There was no degeneration shown in sections of the spinal cord stained by the Weigert hematoxylin method. Sections of the anterior and posterior lumbar roots and of the anterior and posterior cervical roots appeared normal when stained by the acid fuchsin and by the Weigert method. The Betz cells of the paracentral lobule appeared normal when stained by the thionin method. Medulla, pons and cerebellum were normal microscopically, as shown by the hemalum-acid fuchsin and Weigert hematoxylin methods.

CASE 11.—J. B., female, aged 67, peddler, was admitted to the Philadelphia Hospital July 16, 1898; died Sept. 9, 1901.

Diagnosis.—Paralysis agitans. Case of Dr. William G. Spiller.

Clinical History.—Tremor began 20 years before admission to hospital, after being scared several times by robbers entering the house. She had a coarse rhythmical tremor of the head and both hands, but especially the right. Her attitude and gait were characteristic of paralysis agitans.

May 11, 1899: Her legs and feet were swollen; the right more than the left, but there was no edema.

Feb. 19, 1900: She sits crouched in a chair. Saliva dribbles constantly from her mouth. Died of uremia.

Pathologic Examination.—Macroscopically, the appearance of the central nervous system was normal.

Microscopically, sections from the cervical region of the spinal cord showed an apparent increase in the number of blood vessels in the white matter due to a moderate thickening of their walls. Sections from the thoracic and lumbar regions showed the same condition as those from the cervical region. There was no degeneration in the spinal cord detected by the Weigert hematoxylin method nor by the Marchi method for recent degeneration. The spinal nerve roots were normal. The medulla, pons and cerebellum appeared normal when stained by the hemalum-acid fuchsin and Weigert hematoxylin methods. The Betz cells of the paracentral lobule, and the anterior horn cells of the spinal cord, were moderately pigmented, but appeared otherwise normal when stained by the thionin stain.

Sections of a nerve appeared normal when stained by hemalum-acid fuchsin and Weigert stains. Sections of muscle showed a marked atrophy. There was a very marked rounding off of the muscle fibers in many of the bundles with increase of the sarcolemma nuclei and loss of the normal striations. There was an increased amount of interstitial connective tissue but no round-cell infiltration. Muscle spindles were not obtained in the sections. An intramuscular nerve filament was normal.

CASE 12.—F. McG., female, aged 62; no clinical history obtained.

Diagnosis.—Paralysis agitans. Case of Dr. William G. Spiller.

Pathologic Examination.—Macroscopically, the appearance of the central nervous system was normal.

Microscopically, in sections from the cervical region of the spinal cord, the small blood vessels appeared somewhat more numerous in both the gray and white matter than is normal, due to thickening of their walls. The nerve cells in the anterior horns appeared normal for the most part, but some of them presented a glassy, hyaloid appearance. The same was true of the thoracic region and a similar condition was present in the lumbar cord. Sections stained by the Weigert hematoxylin method showed no degeneration of the spinal cord. There was no marked increase of glia in the spinal cord. The spinal canal was open. The pia and the anterior and posterior nerve roots were normal. The medulla and cerebellum were not examined.

CASE 13.—S. H., female, aged 59, was admitted to the Philadelphia Hospital May 7, 1901; died Jan. 27, 1903.

Diagnosis.—Paralysis agitans and rheumatoid arthritis. Case of Dr. William G. Spiller, and was reported by him.

Clinical History.—Her present trouble began two years before admission to the hospital with painful swelling of the knees.

June 21, 1901: There began a course tremor of all parts of the right upper limb partially arrested by voluntary movement. There was no involvement of the cranial nerves. The knees were swollen and crepitated on motion. There was atrophy of both legs, especially the thighs and also of the upper limbs and trunk muscles. Resistance to passive movement was proportionate to the musculature. There was ulnar deviation of the hands. Knee-jerks were much diminished. The back was very rigid. She died in a sudden cyanotic stupor attack.

Pathologic Examination.—Macroscopically, the appearance of the central nervous system was normal.

Microscopically, sections from the paracentral lobules showed a slight thickening of the pia and considerable congestion of the blood vessels. The Betz cells appeared normal when stained with the thionin stain and were not excessively pigmented. Sections from the cervical region of the spinal cord appeared

normal when stained by the hemalum and acid fuchsin and the Weigert hematoxylin stains. The spinal canal was open and there was no proliferation of neuroglia about the blood vessels within the cord. The anterior horn cells appeared normal when stained with the thionin stain. Sections from the thoracic and lumbar regions of the spinal cord showed the same conditions. There was no sclerosis of the blood vessels in the spinal cord. The anterior and posterior spinal nerve roots in the cervical region appeared normal when stained with the hemalum-acid fuchsin and Weigert hematoxylin stains. Sections of a nerve from the arm appeared normal. Sections of a nerve from the leg (it is not known from which leg) showed a partial but decided degeneration when stained by the Weigert method. There was a moderate thickening of the nerve sheath but no round-cell infiltration.

CASE 14.—G. B., male, aged 74.

Diagnosis.—Paralysis agitans (?), cystitis, myocarditis. Case of Dr. J. H. Lloyd.

Clinical History.—Family history was negative and previous illnesses had no bearing on his condition. He denied all venereal disease and the excessive use of alcohol. On admission to the Philadelphia Hospital in May, 1906, he complained of gradually increasing weakness and stiffness in all four extremities and occasional attacks of vertigo. Examination at that time showed a moderately emaciated man, unable to walk or stand. There was no paralysis of the face or extremities. The biceps, triceps and knee-jerks were prompt but about equal on each side. There was no disturbance of sensation anywhere. There was no Babinski reflex and no ankle clonus. There was a marked tremor of the hands, especially of the right, which occasionally stopped momentarily. There were at times fibrillary tremors in the calf muscles. He died suddenly on June 21, 1906. Necropsy was held 75 minutes after death and before the onset of rigor mortis.

Pathologic Diagnosis.—Paralysis agitans, cystitis, myocarditis and ependymitis of the lateral ventricles of the brain.

Pathologic Examination.—The brain weight was 1,140 gm. There was a slight milkiness of the pia of the convexity. The convolutions about the posterior pole of the brain were somewhat smaller than normal and the Rolandic fissure on each side extended into the fissure of Sylvius, otherwise the convexity of the brain was normal. Horizontal section of the brain showed a well-marked ependymitis of the third ventricle and of each lateral ventricle. There was an adhesion completely shutting off the posterior portion of the right lateral ventricle from the main portion, anterior. There was another adhesion which partially occluded the left lateral ventricle in a similar manner. Projecting from the left caudate nucleus into the anterior horn of the lateral ventricle of that side was a pea-sized tumor formation. The choroid plexus was cystic. There was

no internal hydrocephalus and no area of softening. The cerebral peduncles, pons, medulla, cerebellum, and spinal cord and its membranes appeared normal, macroscopically. Microscopically, the tumor formation in the anterior portion of the left lateral ventricle was seen to be surrounded for about two-thirds of its circumference, with an uninterrupted layer of ependymal cells. The cortical portion of the tumor was densely fibrillar, the central portion contained numerous lacunæ and large ependymal cells arranged in alveolar formations.

Sections from the paracentral lobules showed considerable congestion of the blood vessels and a slight thickening of their walls. The pia was slightly thickened but there was no increase in the subpial neuroglia layer. The Betz cells appeared fairly normal by the thionin stain, though a few were observed in which there was evidence of a degenerative change, chromatolysis and displacement of the nucleus to the periphery of the cell. Sections of the cervical region of the spinal cord showed a large number of amyloid bodies, especially in the posterior columns. In some sections here was a well-marked doubling of the spinal canal. There was an increased amount of the neuroglia in the sections, more marked in the posterior columns. The smaller blood vessels and capillaries showed a decided thickening of their walls. The pia was normal and there was only a slight thickening of the walls of the larger blood vessels. In sections from the thoracic and lumbar regions of the spinal cord, the appearances were similar to those in the cervical region. The anterior horn cells in sections from the cervical and the lumbar region stained by the thionin method appeared normal and contained but a small amount of pigment.

The cells of the Clarke's column showed typical chromatolytic changes with displacement of the nucleus, to the periphery of the cell. It is noteworthy that in this case this condition could hardly have been due to postmortem changes as the necropsy was done almost immediately after death. Sections of the spinal cord stained by the Weigert hematoxylin method showed no degeneration. The anterior and posterior nerve roots were congested but appeared otherwise normal. Sections of the median nerves from both arms showed an overgrowth of the sheaths of the nerves and also an overgrowth of the connective tissue within the nerve bundles. The nerve fibers appeared normal and stained well by the Weigert hematoxylin method.

Pieces of muscle from the right and left biceps and the right and left posterior tibial muscles were examined microscopically. The appearances were practically the same in each and may be described as follows: The muscle fibers were somewhat irregular in size. Many of the fibers were thin, tapering and convoluted. These convoluted fibers would often lie between two apparently normal ones. Some of the fibers presented a distinct hyaloid appearance. On cross section the fibers appeared

rounded off and occasionally contained vacuoles. The transverse striations were generally indistinct and in some fibers lost; the longitudinal striations appeared unduly prominent. There is an increased number of nuclei within the fibers, many of which do not have the appearance of typical sarcolemma nuclei, but are small, round or irregular and dark staining. The amount of interstitial tissue is increased somewhat but the rod-shaped nuclei are not frequent. Although sedulously looked for, no muscle spindles were observed. The intramuscular nerve fibers stained well with the Weigert hematoxylin method.

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MORBID SLEEPINESS

WITH A REPORT OF A CASE OF NARCOLEPSY AND A REVIEW
OF SOME RECENT THEORIES OF SLEEP

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MORBID sleepiness occurs in two forms, a continuous drowsiness or sleepiness extending over a prolonged period of time and of various degrees of intensity; and attacks of sleep which vary in depth, from mere drowsiness to complete unconsciousness and in duration from a few minutes to many hours. The continuous form is by far the more common, and for it many causes are assigned:—old age, weakened heart and diseased arteries; diseased vascular conditions preceding cerebral hemorrhage, cerebral malnutrition or inflammation occurring during or before certain forms of insanity: various toxemias, malarial, cholemic, uremic, syphilitic, dyspeptic and diabetic; isolation; cerebral anemia and hyperemia; cerebral tumor and cranial injuries; exhausting diseases, and African sleeping sickness. The above list given by Dana contains certain obvious duplications, but cases are common in which sleepiness, so called, is a symptom of each of these conditions; this sleepiness, however is, in most of the cases, a condition of stupor or coma which obviously differs from true sleep. Skerrit and Stewart report the case of a youth seventeen years old who after a period of hard study and insufficient sleep, slept for fifty days. Marduel reports a case of prolonged sleep associated with a great increase in the excretion of urea. Hysteria, hypnotism, or auto-suggestion is the cause of prolonged sleep in some cases, though in many of these there are demonstrable differences between the lethargic or trance state and normal sleep. Such cases are reported by Blondet, Hayman, John Gay, James Edwards, Drosdow, and others. In this connection there should be noticed the case reported by W. T. Gairdner: A woman thirty-two years old who four weeks after the birth of her child fell into a trance lasting

twenty-one months. She had to be fed with a stomach tube, passed her urine and feces in bed, and cayenne pepper blown up her nose produced no results. She was suddenly cured without therapeusis and the sequel showed it to have been a case of "motiveless malingering."

Of more interest because more rare are those cases of the second form to which I have referred, those in which the sleepiness comes on in periodical attacks. They may be a symptom of many of the conditions which are given as the cause of the continuous drowsiness; they may be symptomatic of anemia (Ward Cousins), or auto-intoxication (Caton); they may occur in association with affections of the liver (Leopold Levi), or be caused by a sarcoma at the base of the brain in the interpeduncular space (Francesco Franceschi); they may be symptomatic of neurasthenia (Oppenheim); and they are not rarely the equivalent of an epileptic attack. In 1880 Gélinau proposed the name "Narcolepsy" for "a rare neurosis characterized by an imperious sleep of sudden onset and short duration which recurred at more or less frequent intervals." The name, derived from the Greek, was suggested because the condition combined the features of somnolence and catalepsy, but Foot objects that "*νάρκη*" implies stiffness, numbness or deadness, and therefore is not descriptive of the actual condition in these cases in which there is simply the quiescence of sleep and suggests, instead "Hypnolepsy," from the Greek *ὑπνος*, meaning sleep. It was not a symptom of any bodily disorder, and Gélinau was decidedly of the opinion that the condition to which he gave the name narcolepsy was not epileptic in its nature or origin, but the name has been indiscriminately used to indicate both the symptomatic and epileptic forms of stupor attacks, which is unjustifiable, as there are symptoms which enable a distinction to be made.

The case which I have to report corresponds to the description by Gélinau of a case of narcolepsy. The patient attended the service of Dr. Wm. G. Spiller at the Polyclinic Hospital in September, 1905.

J. R. G. is a slate quarrier, forty-one years old and married. His father was killed by an accident, but his mother is living and well. There is no history of epilepsy

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or any other nervous disease in the family. One brother and one sister are living and in good health; five others died in infancy or early youth. The patient had scarlet fever when a child and typhoid fever when about twenty-one years old; since then he has been well until his present illness. He has been married twenty-two years; his wife is well, has had four children and no miscarriages; two of the children are living and well; one died of convulsions when two years of age, another died of diphtheria when ten years of age. The patient drinks one glass of beer daily but no whisky and uses tobacco chiefly by chewing.

He was entirely well so far as he knew until the morning of August 25, 1904, when while walking in the street he felt a slight twitching in the legs and they "gave way." On the same day he attended a political convention and while there fell asleep. After sleeping about five minutes he was awakened by a friend but soon afterward fell asleep again; he was reawakened and then went home. Ever since then, until his examination in September, 1905, he has had these attacks of sleepiness coming on at irregular intervals many times a day and persisting until he is awakened by some slight stimulation, such as a spoken word or a slight touch. The attacks bore no relation to taking food, and occurred often in the most inconvenient or perilous places, such as while talking, while being examined in the hospital, while eating, when working in the quarry, ascending ladders, etc. The attacks were preceded by a feeling of depression and fatigue. For a few days after the onset of his illness he had a severe diarrhoea which he attributed to "catching cold." He said that he had become more excitable and irritable and that when excited or angry or even when joking he had involuntary, general, incoördinate movements, would drop objects held in his hands, and his legs would be liable to "give way" under him but he would not fall. When walking and he felt sleepy he would also feel great fatigue in his legs, but if he slept for five minutes this feeling would have left him when he woke. He slept a normal amount at night but dreamed frequently. The dreams were usually of an unpleasant nature; "a woman reaches over and grabs me by the throat but I cannot move," that he was in an asylum

for the self-polluted, etc. He seemed to have a vivid memory of his dreams. He had no hallucinations of sight, but sometimes "imagined people spoke to him when they did not." He was absent-minded, his memory poor for recent events, but he can usually recollect after an effort. He felt well when he arose in the morning and not tired or melancholy. He had no headaches, no indigestion nor any other symptom or complaint. His appetite was good and his bowels moved daily.

Examination showed him to be a well-nourished, muscular man with the appearance of perfect health. He answered questions quickly and intelligently. Several times during the examination, which was conducted in the presence of a number of students, he fell into what was, to all appearances, a sound and peaceful sleep, but from which he could be quickly and easily awakened. His pulse was regular, the rate and tension normal. Examination of his chest and abdomen was negative. His gait and station were normal and there was no paralysis or ataxia of the face or limbs. The tendon reflexes of the upper and lower extremities were prompt and normal on each side; there was no ankle clonus or Babinski reflex. His tongue was clean and there was no tremor or atrophy. Sensation to touch and pin-point was normal all over the body. The palate was equally sensitive on each side and the conjunctival and skin reflexes were normal. The sense of smell seemed slightly more acute in the right nostril. The sense of taste was normal and equal on each side of the tongue. No areas of tenderness or hysterogenous zones could be detected. An eye examination made by Dr. Hansell was negative; the fundus oculi, the pupillary reflexes and the visual form and color fields being normal. The urine contained no albumen or sugar. A blood examination was not made, but the conjunctiva and mucous membranes were of good color.

To summarize:— a strong, somatically healthy, laboring man forty-one years old suddenly begins having attacks of sleep many times a day, evidently involuntary but unaccompanied by any of the physical signs of illness, even the stigmata of hysteria being absent. There is a slight mental change, the heightened irritability being perhaps not unnatu-

ral under the circumstances. His natural sleeping period is undisturbed except by particularly vivid and unpleasant dreams.

The administration of caffeine was of no use in keeping him awake, and his principal treatment consisted in the application of static sparks to his head with suggestion. This with general hygienic treatment and the change of scene and occupation appeared to be of benefit as in a week's time the number of attacks daily was considerably diminished.

Gélineau quotes Caffé as being the first to report a case of this description. These cases must be distinguished from epileptic pseudo-narcolepsy and from hysterical pseudo-narcolepsy; the differential diagnosis will be referred to later. Genuine cases of narcolepsy are not common in the literature. MacNamara, in 1862, published the case of a girl, fifteen years old, who was attacked with severe pain in the head which was worse when she was in the recumbent position. At the same time she began having sudden attacks of sleep while eating, at the piano, coming down stairs, etc., from which she could be awakened at the slightest touch. She was entirely cured by a severe epistaxis. E. Mendel reported a case in a man, aged thirty-seven years, who also had migrainous attacks, and the author thought that both were due to an affection of the sympathetic nervous system leading to cerebral anemia. Matas coincides with this view of the affection. Morton reported a case in a physician, thirty-two years old, who went to sleep on the slightest mental exertion, such as reading, while making a vaginal examination, three times during the writing of a prescription, etc. He was utterly unable, when taken off his guard, to control his temper, and he had such a dread of mental exercise that he would put off settling a question, even of a trivial nature, rather than think about it. Legrand reported a case in a girl, aged sixteen years, who also had a facial tic. The attacks of sleep only lasted from twenty to sixty seconds and the awakening was spontaneous with a profound inspiration. Jacoby reported a case in a barber who had no other symptoms except that he was gaining fast in weight. Franz Fischer reported a similar case as epileptoid, though the

details given are against such a diagnosis; a single woman, twenty-two years old, would suddenly fall asleep from two to six times daily whether at work or idle; just before falling asleep she would feel very tired, but whether the sleep lasted five minutes or an hour she awoke feeling refreshed and was immediately able to continue her work; she had insomnia and unpleasant dreams, but all of her bodily functions were normal, and treatment by strong galvanism to the head greatly alleviated her condition. Foot reported a case in a patient, eighteen years old, of neurotic heredity who went to sleep every day at three p.m. no matter what he was doing. The attacks came on gradually but he was powerless to resist them. There was no convulsion or spasm, and the author does not regard the condition as epileptic. In this case the inhalation of amyl nitrite, or the administration of picrotoxin or caffeine were of no benefit. Ewen reported a case in a soldier, twenty-four years old, who was arrested for sleeping on guard duty. He often slept through roll-call and had been frequently punished for it. On one occasion he was found asleep on a ladder fifteen feet from the ground, at another time, at a meal, with a soup-spoon in his mouth. He appeared to be unconscious of the fact that he had been asleep or that he slept more than other men. There were no signs of epilepsy, hysteria, or cretinism. Chavigny reported an almost identical case also in a soldier; the only somatic disturbance being a slow pulse, forty to fifty per minute. In a case reported by MacCormac the apparent exciting cause of the condition was the extraction of ten teeth without an anesthetic. The attacks would last from three to ten minutes or longer and the patient would have twelve or more daily. She was entirely unable to prevent their onset and they came on under the most varied circumstances, though she was easily awakened. She was treated with phosphate of iron, quinine sulph., tinct. nux vomica and nitro-glycerine, also galvanism to the head, a weak current, three minutes at a time, three or four times weekly. She was cured, but the condition recurred as a result of worry. Dana, D. J. McCarthy and recently Friedmann have reported similar cases but also include as narcolepsy cases of a *petit mal* and hysterical somnolence.

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Cases of hysterical sleep attacks are numerous in the literature, and are referred to by Armaingaud, Legrand, Achard, Gilles de la Tourette, Pitres, P. Richer, Charcot, Parmentier, and others. Camuset's patient had a dracocystitis and every time a sound was passed up his lachrymal duct he went into a sleep lasting from six minutes to an half hour from which he awakened perfectly normal; any emotion provoked an attack. Gilles de la Tourette regarded the sleep attacks which occur in the early stages of paresis as due to a superimposed hysterical state, and inasmuch as the hysteria tends to disappear in the course of the disease it explains the frequency of the attacks in the earlier stages. Cases of hysterical pseudo-narcolepsy can be fairly easily diagnosed clinically by certain features of the attacks themselves, such as the vibratory movements of the eyelids, a certain stiffness of the extremities, position of the body, etc.; also by the cause of the attacks, suggestion, and the mode of onset. They are in reality hysterical trance states and present at least some of the well-known features of this neurosis; the presence of the stigmata of hysteria, sensory changes, hysterogenous zones, etc., will confirm the diagnosis.

The sudden unconsciousness of attacks of *petit mal* are too well-known to need extended notice here. Ch. Féré has called attention to attacks of somnolence occurring in epileptics, and at times replacing the epileptic attacks, which are characterized by suddenness of onset and complete unconsciousness from which the patient cannot be awakened though he may be, in appearance, only asleep. In another place the same author calls attention to attacks of "epileptic apathy" differing from post-epileptic depression, and resembling attacks of bromism, from which it is important to distinguish them. This may be done by the absence of gastric and respiratory derangement and their cure by the continuation of the bromides in larger doses. These forms of epileptic manifestations are obviously different from the attacks of narcolepsy described by Gélineau, of which the case herein reported is an example. There are reported cases in which the basis of the sleep attacks were either hysterical or epileptic but in which the signs

of both diseases were combined in such a way that their differentiation was difficult, as in the cases reported by Sahlman, Rousseau, Porter and others. The cases of Friedmann, while he refers to them as "not epileptic," evidently bore a very close relation to that neurosis; in several the attacks occurred in patients who had, or later developed, genuine epileptic attacks; they were in some cases attended with relaxation of the sphincters, etc., and the description of the attacks themselves, the sudden onset, staring eyes, lack of response to stimuli, etc., show their epileptic nature.

Oppenheim, in his text-book, and Ch. Féré in a paper on the subject "*Le sommeil paraxystique*" regard narcolepsy as merely a symptom, and taken simply in the sense of paroxysmal sleep attacks this is undoubtedly correct. Oppenheim speaks of them under the headings of epilepsy, hysteria, and neurasthenia, and Féré also includes various organic causes. It is true that sleep attacks are symptomatic of these affections, but it is also true that sleep attacks that are different from the symptomatic attacks described by these authorities are occasionally met with, and it is to these that the name narcolepsy or some similar name should be given. Ballet in his "*Contribution à l'étude du sommeil pathologique (Quelques cas de narcolepsie)*" refers to narcolepsy as merely a symptom, the opposite of insomnia, for which he gives as causes: First, visceral diseases and disturbed nutrition; second, vicious hygiene, eating too much, etc.; third, deficient oxidation, diabetes, etc.; and fourth, vicious functioning of the nervous system. All nervous diseases, in a sense, are due to impaired or vicious functioning of the nervous system; epilepsy, hysteria, and many others may be included in this category; why not then make of these cases of narcolepsy a separate disease or at least a syndrome? They have sufficient individuality to permit of their being readily diagnosed from other neuroses or from the merely symptomatic attacks of sleepiness; they are not of themselves symptomatic except of neuro-psychic degeneration (Ribakoff) which is a reputation they would share with the other neuroses so-called; and finally they are benefited by a distinctive treatment. I agree with Foot

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that the name hypnolepsy is a much better descriptive term than narcolepsy, and its adoption would tend to lessen the confusion on this subject. It is best described as a syndrome, occurring in individuals with at least a functionally degenerated nervous system, and consisting of irresistible attacks of sleep, ariable in frequency and regularity, irrespective of occupation, surroundings, or of the taking of food or other somatic condition and from which the patient can be easily aroused with the full possession of his mental faculties.

Similar sleep attacks are but rarely a symptom of organic or visceral diseases, and their diagnosis presents little difficulty. The diagnostic features of hysterical pseudo-narcolepsy have already been given. Epileptic pseudo-narcolepsy is distinguished by the sudden onset, the stiffness and rigidity of the limbs, staring eyes, etc., which are not the appearances of sleep, the difficulty of awakening, and the post-epileptic confusion.

What effect has the existence of such cases as these on our ideas concerning the philosophy and the physiology of sleep, and what, in particular, is the cause of these peculiar sleep attacks?

The man whose history is given in this paper when in one of his sleep attacks is to all signs and appearances enjoying a peaceful, natural sleep. Sleep, like food is necessary to the continuation of the life and health of the individual, but we do not ordinarily eat or sleep on this account. Reproduction is necessary to a continuation of the species, but most people will admit that sexual desire is the impelling force which is acting on the individual. In a like manner it is hunger, the desire for food, that impels us to eat, and there is a sensation, in many respects similar to these, that impels us to sleep. The desire to sleep, like the desire to eat, is usually regarded as indicative of an organic need, but neither is necessarily so. In many organic diseases, especially those affecting the stomach, the desire to eat may be much increased. Gluttonous individuals who eat enormous quantities of food for which there is no organic need are not uncommon; in various mental diseases gluttony is a symptom, and finally there are reported cases of individuals in

whom hunger recurred at such short intervals and was so imperious that the patient had always to carry some article of food with him. Cases of the sort last referred to with a perversion of the appetite for food correspond closely with the case reported in this paper in which there was a perversion of the appetite for sleep. Both conditions must be regarded as due to a mental defect. The time of sleeping and the amount that will be taken is rather a habit than a fixed rule of nature and is capable of the widest individual variations. The average individual sleeps a certain portion of each twenty-four hours; it is his fixed habit to do so, and at a certain time each day he feels sleepy, but a perversion of the desire to sleep, a perversion of a habit, will bring on the desire to sleep at any time and with added intensity. Such an explanation of the case offers no difficulties to those who have given any study to organic desires in general and their perversions.

Pierre Janet has analysed the pathogenesis of some of these impulsions, among which is the case referred to above, in which the patient at short intervals is imperatively attacked with the desire to eat. As a result of the psychological analysis of these cases he concludes that the impulsions arise in neuro-paths as a result of attacks of depression from emotional causes and to these the impulsions are joined in a rather accidental manner. For instance, in the above case of Janet's the patient first feels weak, depressed, empty, etc., for the relief of which she takes food; the taking of food soon becoming an imperative necessity. The manner in which the impulsion to sleep arises in such cases as the one reported in this paper is in all probability identical with the above. The patient feels depressed, weak, tired; if walking "his legs give way under him" just before the sleep attack; a natural mental process indicates sleep as a remedy upon which basis sleep becomes an imperative idea. An indication that the tiredness he feels is not real fatigue is found in the fact that no matter how short the time he sleeps he awakes feeling refreshed and strong. The condition is not hysterical; Janet makes the statement: "We must not forget that the cause of the impulsion is, above all, in the underlying attack of depression; we must endeavor

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to discover, what is unfortunately not always possible, the physical and moral conditions that determine it; and we must by all sorts of hygienic means, by proper mental treatment, prevent its reappearance. It will be easily seen that the therapeutic measures would be different in cases where the impulsions depend on sub-conscious phenomena or suggestions as is often the case among hysterics."

It is obvious that this view of the pathogenesis of the impulsion as applied to attacks of sleep is not consistent with many of the older theories as to the production of sleep such as fatigue, auto-intoxication, etc., but makes sleep a positive function of the mind; so that from a study of this case we arrive by another route at the same conclusion as Ed. Claparède that "sleep is not a purely negative or passive state, not the result of an arrest of function: it is a positive function, an act of a reflex order, an instinct." This instinct may be regarded as developing along well-recognized lines of evolution, according to the law of "the survival of the fittest," and in a plane analogous to the instinct of self-preservation. According to this theory Claparède does not regard sleep as the result of fatigue or auto-intoxication but as a reflex designed in normal persons as a protection against fatigue and its consequences. "The mechanism of sleep consists of a reaction of disinterest for the situation present. It is not the irritability or the receptivity which is abolished, but the reactivity of interest and the reactivity of adaptation. The organism profits by the arrest of muscular effort, and the relaxation of the 'mental tension' is probably compensated for by an increase in the vegetative tension." This "biological theory" of sleep explains in a much more satisfactory manner than any other theory the correlated phenomena of sleep; for instance, the onset of sleep may be prevented by the presence of more powerful instincts or those which have, at the time, a greater interest, such as the instinct of self-preservation; a fact which it would be difficult to explain if sleep were due to fatigue or the presence of toxic substances in the body. In man and in many animals sleep follows a natural physiologic rhythm; this can be explained in the same way that Claparède explains hibernation; that it is a secondary adaptation to environment.

Claparède reviews the theories that have been suggested for the cause of sleep: cerebral anemia and hyperemia; lymphatic hypertension, which is similar to the osmotic theory recently put forward by Devaux; interruption of conductivity in various parts of the brain or at the periphery; the histologic theories, retraction of the dendrites, etc.; the inhibition of intellect; default in excitations; and the supra-physiologic hypotheses; and finds as objections to all of them, that they are based on uncertain data; that they are in many cases more likely a consequence than a cause of sleep; and finally, supposing that these phenomena were the cause of sleep, their cause would still require explanation. The bio-chemical and toxic theories are open to the objections that:—sleep and fatigue are not parallel conditions; sleep can be voluntarily retarded and is retarded by instincts of greater interest; the influence of quiet, or of monotonous stimulation, such as a sermon, in producing sleep; and the partial sleep from which the sleeper is aroused instantly by a particular stimulus while sleeping soundly through other stimuli; for instance, the mother who wakens at the first cry of her baby.

A. Lorand regards sleep as a function of the mind but insists that it is regulated by the secretion of the thyroid gland. The evidence that he brings forward to support this assertion is far from conclusive, and the same may be said of Alberto Salmon's hypothesis that the internal secretion of the pituitary body is the cause of sleep.

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THE SYMPTOMATOLOGY OF LESIONS OF THE LENTICULAR
ZONE WITH SOME DISCUSSION OF THE PATHOLOGY
OF APHASIA.*

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INTRODUCTION.

The views on aphasia recently advanced by Marie have served, among other things, to concentrate the attention of physiologists and neurologists upon the functions of the lenticular nucleus, to which body Marie has given an unusual role in the production of motor aphasia. It is a noteworthy fact that, in spite of the neurological activity all over the scientific world during the last three or four decades, the functions of the basal ganglia still remain largely undecided. Many speculations have been advanced regarding the functions of the thalamus, caudatum and lenticula; and many facts, anatomical, physiological and clinicopathological, have been contributed regarding these organs; but the comparative certainty of our knowledge as to the functions of the cerebellum, most of the cortical fields and centers, and many of the encephalic tracts of projection and association, is lacking with regard to these ganglia. The reasons for this state of affairs are not hard to see. Owing to the deeply seated position of the basal ganglia, it is difficult to experiment on them without involving or injuring other parts; and the lesions which are observed by the clinicopathologist are rarely absolutely limited to any one of these organs. Microscopical investigations of degenerations of these ganglia and of their incoming and outgoing tracts have not led to as clear inferences as in other regions of the brain.

Our paper is based in the first place upon a study of eleven cases with necropsies, in ten of which the lesion involved the

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lenticular zone. In six of these cases the lenticula itself was involved. Other parts of the lenticular zone were variously implicated in the ten cases. It was possible, however, at least in some of the cases, in which regions other than the lenticula were involved, to arrive at an idea as to the part played by this body in the symptomatology presented by the patients.

For several years the writers of the paper have been collecting cases with necropsies in which lesions of the basal ganglia have been present. Most of the specimens from these cases are to be found in the Laboratory of Neuropathology of the University of Pennsylvania. In not a few instances papers have been published in which references have been made to some of these cases, and some use will be made of these published but personal data. In going over the specimens of cases, clinical and pathological studies of which have not yet been made, the writers found that they had in their possession more than twice as many specimens of cases as have been used in this paper, but owing to the imperfections of the clinical data, or because of the confusing character of the lesions, only eleven of these cases could be used with any advantage.

OLDER VIEWS REGARDING THE FUNCTIONS OF THE LENTICULA.

We shall first note briefly the views regarding the functions of the basal ganglia held by some of the older physiological and neurological teachers.

Fifty years ago one of the most relied-upon general works in physiology was that of Carpenter.¹

An old view which is discussed but antagonized by him was that the thalami in some way were centers for movements of the upper extremity, and the striata for the lower. Carpenter also opposes the idea of Magendie that removal of the striata causes an irresistible tendency to forward progress, whilst the division of the peduncles of the cerebellum occasions the reverse movement.

Discussing the functions of the striatum, Carpenter says that, "According to Longet, Schiff and Lafargue, the results of re-

¹"Principles of Human Physiology," by William G. Carpenter, M.D., F.R.S., F.G.S., Examiner in Physiology and Comparative Anatomy in the University of London, etc., fifth American from the fourth and enlarged London edition, Philadelphia, 1853.

removal of the corpora striata with the anterior upper part of the cerebral hemispheres, are for the most part negative; for the animal usually remains in a state of perfect stupor, although still retaining the erect position; and it is only when irritated by pinching or pricking that it will execute any rapid movements. No mechanical irritation of the corpora striata produces either sense of pain or muscular movements. No distinct evidences regarding the special functions of either of these ganglionic masses can be gained from pathological phenomena. So far as is yet known extensive disease of either the thalami optici or the corpora striata of one side, produces hemiplegia or paralysis both of sensation and motion on the opposite side. The same result very commonly follows an apoplectic effusion into the substance of either."

Broadbent,² writing in 1876, voiced the views which were held by many about the time and shortly after the time when the study of cerebral localization received its greatest impetus through the suggestions of Hughlings Jackson (1864) and the researches of Hitzig, Fritsch, Ferrier and others (1870-1875).

Discussing the position of the thalamus and striate bodies, with reference to the crusta and tegmentum, after indicating the existence of connections between these ganglia and the other parts of the nervous system, both lower and higher, he expresses the opinion that the striatum is a motor and the thalamus a sensory ganglion for the entire opposite half of the body. With regard to the striatum, like all the earlier writers not separating in his discussion the caudatum and the lenticula, Broadbent holds that this ganglion translates volitions into actions and puts into execution the commands of the intellect. "It selects, so to speak, the motor nerve nuclei in the medulla and cord appropriate for the performance of the desired action, and sends down the impulses which set them in motion. These impulses are transmitted through fibers, and the fibers must start from cell processes in the corpus striatum. A given movement, therefore, must be represented in the corpus striatum by a group or groups of cells giving off downward processes, which become fibers of the motor tract of the cord." When the movement is simple the cell group will be small and the fibers few; when complex the cell group

²Broadbent, Wm. H. *British Medical Journal*, April 1, 1876.

will be large and well defined and the descending fibers numerous.

"Words," says Broadbent, "which require for their utterance the simultaneous co-operation of the muscles of the chest, larynx, tongue, lips, etc., and the exquisite and rapid adjustment of their movements concerned in phonation and articulation, must be represented in the corpus striatum by very large groups of cells, and not in that of one side only but in both."

According to Ferrier³ it is evident that destruction of the striatum produces a much more complete and enduring paralysis than destruction of the cortical motor centers alone, or of the pyramidal tracts which proceed from them.

He summarizes his views with regard to the corpora striata as follows: "It appears from these facts that the corpora striata proper are centers of innervation of the same movements as are differentiated in the cortical motor centers, but of a lower grade of specialization. The innervation of the limbs in all that relates to their employment of instruments of consciously discriminated acts is dependent on the cortical centers, while for all other purposes involving mere strength or automatism, primary or secondary, the corpora striata with the lower ganglia are sufficient. In man almost every movement has to be elaborately acquired by conscious effort through the agency of the cortical centers, and continues to involve the activity of these centers to a greater or less extent throughout. Hence the destruction of the ganglia of the corpora striata adds little if anything to the completeness of the paralysis which results from destruction of the cortical motor centers alone."

Ross⁴ has contributed something to our more exact knowledge of the motor functions of the lenticula in a paper on what is now commonly designated as pseudo-bulbar paralysis, the title of his contribution, however, being labio-glosso-pharyngeal paralysis of cerebral origin. He cites a case from Oulmont⁵ with the typical symptoms of pseudo-bulbar paralysis in which the motor and speech phenomena were apparently due to bilateral lesion of the lenticula.

³"The Functions of the Brain," by David Ferrier, M.D., second edition, rewritten and enlarged, New York, 1886.

⁴Ross, James. *Brain*, July, 1882.

⁵Oulmont. Cited by Lépine, R. *Mensuelle de Médecine et de Chirurgie*, Vol. 1, 1877.

"An old hemorrhagic focus was found in the right hemisphere occupying the third or external segment, and a smaller focus situated in the second segment of the lenticular nucleus. Similar foci were found in the third and second segments of the lenticular nucleus of the left hemisphere. The internal capsule was not involved in either hemisphere. The medulla oblongata and pons were free from disease."

Ross cites a case of Eisenlohr⁶ in which lesions were found in both striata (the lenticular nuclei?) and in the thalami.

Ross also cites the case of Kirchhoff,⁷ published in 1881, in which lesions of both the caudatum and lenticula were found, but as the capsule also seems to have been involved in this case, it is not of as much value for our purposes as other cases in which the lenticula alone seems to have been implicated.

One of the most important cases of labio-glosso-pharyngeal paralysis or pseudo-bulbar paralysis from disease of the lenticula which has as yet been described is that reported by Ross himself. In this case the lesions described were as follows:

"The lenticular nucleus of each hemisphere was found post-mortem replaced by a well-defined cystic cavity, containing a clear, straw-colored fluid, the internal capsules being apparently uninjured. Microscopic examination failed to detect any evidence of disease in the nerve nuclei of the medulla oblongata, or descending changes in the pyramidal tracts in any part of their course."

In another case recorded by Ross the lesions present were described as follows:

"The anterior part of the lenticular nucleus of the left hemisphere presented two small cyst-like cavities, containing clear fluid. These cavities occupied the second and third segments of the nucleus, and one of them appeared to encroach to some extent on the knee of the internal capsule. The right hemisphere was preserved in spirit, and was not examined until a fortnight later, when a small cavity was found in the lenticular nucleus of this hemisphere also. This cavity occupied the anterior part of the nucleus, but did not appear to encroach upon the internal capsule." Some descending degeneration, probably due to lesion in the internal capsule, was present in this case.

⁶Eisenlohr, C. *Archiv. für Psychiatrie*, Vol. IX, 1878, p. 43.

⁷Kirchhoff. *Archiv. für Psychiatrie*, Vol. XI, 1881, p. 132.

In a third case recorded by Ross a small cystic cavity was found in the left lenticular nucleus close to the border of the genu of the internal capsule. No secondary degeneration detectable by the naked eye was in the crura. A careful microscopical examination of successive sections of the crura, pons, and medulla oblongata, failed to detect any secondary degeneration of the pyramidal tracts. The bulbar nuclei were healthy. This case, which was complicated with disease of the spinal cord, presented typical pseudo-bulbar paralytic symptoms.

Ross held that his cases did not prove that the lenticula is an independent center for the regulation of all movements of articulation and deglutition. He was also inclined to negative the view that the lenticula is a ganglion of interruption between the cortex and the parts below, and looked with favor on the idea that in the cases of labio-glosso-pharyngeal paralysis recorded by him the symptoms were due to lesions interfering with the paths of conduction from the cortex, although at least two of his three cases seem to have been instances of lesions isolated to the lenticula, these causing disorders of speech and some paresis of the face and upper extremity. His view would appear to be in a general way that of Dejerine that the cases of pseudo-bulbar paralysis attributed to lesions of the lenticula, either unilateral or bilateral, are probably dependent upon undetected lesions of the pyramidal tracts or cortex.

THE VIEWS OF DEJERINE.

In his *Anatomy of the Central Nervous System*⁸ Dejerine in several places goes rather fully into the question of the functions of the lenticula, but his results would seem to be included in the assertion that this ganglion plays only a negative part. He is mostly concerned with denying its connection with the motor and other portions of the cortex by projection fibers, appearing to hold that the lenticula plays no part in the control of movement or in the function of speech. In order that his views with regard to the functions of the striate bodies may be understood, it will be necessary to include a reference to his opinions as to the anterior limb of the internal capsule. Lesions of the anterior seg-

⁸Dejerine, J. "Anatomie des centres nerveux," Vol. II., fasc. 1, Paris, 1901.

ment of the internal capsule, says Dejerine, are not manifested by any appreciable motor disturbances when they occupy only the anterior half or three-quarters of this segment. These fibers are, he believes, frontothalamic projection fibers. He says that as soon as the knee of the capsule is reached, a distinct symptomatology appears,—that of paralytic phenomena on the opposite side of the body. Secondary degeneration of this part of the capsule is also seen following cortical lesions of the Rolandic zone. Dejerine has shown that the area of degeneration in the anterior portion of the posterior limb of the capsule is correspondingly more posterior, according as the lesion is higher and higher in the Rolandic cortex, his own observations confirming in this respect the physiological observations of Horsley and Beever on the internal capsule of the monkey.

When the knee and the adjacent portion of the posterior limb of the internal capsule are involved in a lesion, a paralysis of the inferior facial nerve and of the half of the tongue on the opposite side is observed. The fibers arising from the frontal operculum and Rolandic operculum pass through the knee and the adjacent portion of the posterior segment of the capsule. Facial paralysis of capsular origin presents the same features as that of cortical origin, the superior facial is not entirely intact.

When the lesion is bilateral and symmetrical, the clinical picture will be that of pseudo-bulbar palsy. The anatomical localization of the paralysis has been and still is differently interpreted. It is known that this may be of cortical origin (bilateral lesion of the Rolandic operculum), subcortical, capsular, pontile, or bulbar, situated in the course of fibers arising in the motor zone which control the nuclei of the motor cranial nerves. Dejerine does not accept the localization of those who have their views based upon cases in which the lesions were supposed to be limited to the lenticula. In one place he says emphatically that the lenticular nucleus does not send fibers into the foot of the cerebral peduncle and does not receive any from the cortex. Further on in the same work, however, he modifies this statement by saying that the *putamen* does not receive any fibers from the cortex, but that the globus pallidus may do so. The lenticular fibers of projection, he holds, belong to the system of strio-thalamic and sub-thalamic radiations.

Dejerine states that symmetrical lesions (hemorrhage, soften-

ing, either in the putamen or in the lenticular nucleus as a whole) are frequently observed in the necropsies of the aged, who have not presented any symptoms of pseudo-bulbar paralysis during life. He recalls the fact that Oppenheim and Siemerling had shown in 1886 that when they observed a bulbar palsy during life in these cases of symmetrical lesions of the lenticula, there were always also foci of softening in the pons and oblongata. According to Dejerine in all the cases in which there was absence of all pontile or bulbar lesions appreciable by the microscope, the pseudo-bulbar palsy seems to have resulted not from the localization described as lenticular by some; but when these cases are studied by the method of serial microscopical sections, the bilateral and symmetrical lesions of the lenticula are found always to have involved the knee and a portion of the posterior limb of the internal capsule.

He speaks of tracing clearly in the retro-lenticular portion of the internal capsule fibers which traverse the globus pallidus. He says that these may appear to be cortico-lenticular fibers, but suggests that inasmuch as they cannot be followed in the corona radiata, except immediately above the retro-lenticular segment, they may rather be lenticulo-caudate fibers, that is caudate fibers of projection or termination.

Dejerine asserts that in cortical lesions, either extensive or confined to small areas, when the material is treated by the method of Weigert and Pal, the globus pallidus does not appear to undergo any modification, either in the form of atrophy or disappearance of fibers, similar to that which is seen in the thalamus in the same case. He acknowledges, however, that we cannot assert that the globus pallidus possesses in such cases as many radiating fibers, or as dense a network of fibers as in the normal state.

It is further stated by him that in two cases of recent cerebral disease, in one of which the lesion was extensive and in the other limited, examined by the method of Marchi, the globus pallidus and the corpus Luys both received fibers of projection from the cerebral cortex.

In still another place Dejerine states that the striatum, especially the caudate nucleus and the putamen, at least in man, do not receive direct cortico-striate fibers; that they receive only cortical fibers of projection traversing them on their way to the

internal capsule (cortico-thalamic, cortico-pontile, cortico-bulbar, cortico-spinal fibers). It seems to be well demonstrated, according to Dejerine, that cortical lesions, whatever may be their situation, their extent, and their duration, do not affect the caudatum and the putamen as they do the thalamus. This contrast existing between the striatum and the thalamus, says Dejerine, was well recognized by Gudden in 1872 as the result of experimental observations. Other well known investigators cited by him have observed atrophy of the striatum following cortical ablation experiments, but this atrophy results probably from the degeneration of fibers of passage and collaterals or injured cortical neurones, and is not comparable with the atrophy, even extreme, in the thalamus under the same circumstances. The very slight diminution in size of the striatum sometimes seen in man following old lesions of early childhood is, in Dejerine's opinion, the analogue of the diminution in volume seen in all these cases in the anterior corresponding half of the encephalon, in the anterior horn of the opposite side of the cord, and in the opposite cerebellar hemisphere. It results as much from degeneration of the collaterals which the system of cortical projection fibers sends to the striatum and the region of the tegmentum, as from the functional inactivity of this region. Dejerine⁹ has expressed his views regarding the lenticle also in a recent paper.

Some others, (Schwalbe, Edinger, Sachs, Bechterew, Marinesco), Dejerine remarks, deny the existence of cortico-striate fibers of projection, but depending on the homology of development of the striatum and of the cerebral cortex established by Wernicke, attempt to prove the existence of a cortico-striate fasciculus of association, and as such describe the occipito-frontal fasciculus and colossal fasciculus of Muratoff.

THE VIEWS OF MINGAZZINI.

In the appendix to his monograph, Mingazzini¹⁰ discusses and antagonizes the opinions of Dejerine.

The arguments of Dejerine, says Mingazzini, appear far from decisively proving that the lenticular nucleus does not possess

⁹Dejerine J. *L'Encephale*, No. 5, May, 1907.

¹⁰Mingazzini, Giovanni. "Sulla sintomatologia delle lesioni del nucleo lenticolare." 1902.

motor fibers, and that therefore a lesion of the same is incapable of producing paralytic disturbance. The argument that the secondary degenerations show that the lenticula does not send fibers into the foot of the cerebral peduncle and does not receive any from the cortex, and its fibers of projection must belong only to the system of the strio-thalamic and sub-thalamic radiations, according to Mingazzini, has little value, since he (Mingazzini) believes that he has shown in the beginning of his monograph that lesions of the lenticula, according to the points at which they occur, seem to give rise to a diverse symptomatology.

Mingazzini believes that it is not true that in every case in which lesion of the lenticula produces permanent disturbances, there must be existing concealed destruction of the internal capsule. He believes that the arguments of Dejerine might just as well be turned around; in other words, it is just as likely that lesions of the lenticula might be present and escape detection in those cases in which capsular lesions are discovered as the reverse. The arguments of Dejerine that because lesions of the pons and bulb are frequently found in pseudo-bulbar paralysis, therefore these are probably present and the real cause of the paralytic phenomena in cases apparently due to lesions of the lenticula, is easily overcome, according to Mingazzini, if we remember that the symptomatology of cerebro-pontile pseudo-bulbar paralysis is much richer than pseudo-bulbar paralysis of pure cerebral origin; in other words the picture of the two diseases is different.

Mingazzini insists on the point that only certain parts of the lenticula transmit motor and sensory fibers or have motor or sensory functions, and it is therefore easy to understand how lesions may sometimes be found in these ganglia in the old or in others without motor symptoms.

Mingazzini does not accept the opinion of Dejerine with regard to the cases collected from different authors, in which atrophy of the lenticula occurred after lesions or arrests of the cerebral lobes or convolutions, but believes that these cases show that anatomical and physiological connection exists between some part of the cortex and the lenticula. Mingazzini lays particular stress upon the researches of Dejerine, already referred to in our analysis of Dejerine's views, in which he found degenerated fibers in the globus pallidus as the result of cortical lesions, since Min-

gazzini says it shows that these ganglia receive fibers of projection from the cerebral cortex.

Leaving his criticisms of Dejerine, let us turn now to Mingazzini's positive statements of his own views based upon personal clinico-pathological observations, his pathological findings being both gross and microscopic. In his monograph he first calls attention to the manner in which the question of the function of the lenticula has been largely passed by, or has been altogether slighted by writers on neuropathology and clinical neurology. He refers particularly in this connection to well known authors like Gowers, Oppenheim, von Monakow, and Brissaud. Unlike Dejerine, who attacks the problem of the functions of the lenticula in his great work on the anatomy of the nervous system, with the result of negating its functional importance, the other authors referred to by Mingazzini have contented themselves with slight or imperfect allusions to the problem of lenticular disease.

Mingazzini, in this monograph, gives details of nine cases of lenticular lesion with necropsies observed by him, and refers to other cases in a preceding publication. We have gone carefully over the notes of these cases, and also the table in which the topography of the lesion and the disorders of articulation and of motility in the face and limbs are summarized. These cases would seem to leave little doubt of the correctness of the conclusions drawn by Mingazzini. One criticism is worth mentioning, however, regarding some of them. Several of these patients were quite advanced in years; some over seventy and others between sixty and seventy. In several of them also abnormal conditions of the meninges and cortex or subcortex were present, as opacities of the membranes, adhesions with decortications, shrinkage of the convolutions, and in one or two instances, actual softening. Making, however, due allowance for these findings and their possible bearing upon the symptoms in limbs, face and speech, we believe that the most important of Mingazzini's conclusions are to be accepted. We shall give these somewhat at length, translating and summarizing his remarks, omitting quotations, as in order to save space we shall not follow his language consecutively and with exactness.

A focus of disease, even of small size, involving only the lenticular nucleus, according to Mingazzini, never fails to mani-

fest itself with motor disturbances. Usually these are dissociated or total paralyses, to which are sometimes added irritative symptoms.

Facial paralysis is very rarely of lenticular origin. Mingazzini has not observed any case of it. He refers to the one recorded by Ross, to which we have already directed attention. Cases of unilateral facio-lingual paralysis and of bilateral facio-lingual paralysis such as is frequently seen in pseudo-bulbar paralysis without involvement of the limbs are also rare. Mingazzini gives three instances of what he regards as facio-lingual paralysis of lenticular origin. In the first the putamen on the left and in the second the putamen on the right in the neighborhood of the genu was softened; in a third a focus of softening was present in the globus pallidus. Mingazzini refers in this connection to the cases of Lépine and Halipre¹¹ which with others have been criticised by Dejerine.

Mingazzini refers to a form of paralysis intermediate between the pure facial and the brachio-facial paralysis which he had previously reported. The patient presenting this symptom-complex, besides having paralysis of the inferior portion of the right facial, was affected with athetoid movement of both hands. Many lacunar foci were found in the left putamen.

A single example of pure brachio-facial paralysis is recorded. The case was that of a patient affected with paresis of the seventh nerve and of the superior limb of the right side following softening of the anterior portion of the left lenticle.

Facio-crural paresis is very rare indeed. Mingazzini says that he has never seen a pure case of this dissociated paralysis, but refers to a case previously recorded by him in which was present oscillatory tremor of the left superior limb with paresis of the left inferior facial distribution and the left lower extremity. Two foci of loss of substance as large as a millet seed were found; one in the external part of the right putamen and the other in the external member of the globus pallidus. In two cases Mingazzini has observed an irritative paretic disorder of the upper extremity from partial destruction of the lenticular nucleus on the opposite side. In one of these cases the limb affected was spastic; in the other it was the seat of clonic move-

¹¹Lépine, R. Op. cit.

ments. In another case in which softening was found in the middle part of the left putamen, the patient was seized occasionally with clonic movements of the hand and the forearm of the opposite side.

While the forms of hemiplegia, with or without irritative phenomena, are sometimes present in lesions limited to the lenticle, the more frequent syndrome, according to Mingazzini, is motor paresis of the entire half of the body. He records five such cases in the monograph we are analyzing and refers to others previously recorded. The paralytic disturbances often pass without being observed by the patient, says Mingazzini. They apply to the physician complaining of paresthesia, he continues, or of pain along the paretic limbs, or else of vertiginous conditions, or weakness of memory, symptoms evidently due to a diffuse cerebral arterial sclerosis. It is only a minute objective examination, he says, which will reveal certain hemiparesis. While the attack of paralysis may be ushered in by an apoplectic stroke with unconsciousness, the symptoms accompanying this are not so severe and persistent as in ordinary cerebral hemorrhage. The patients are often syphilitic, and probably because of this the attacks are frequently preceded by headache and febrile symptoms. As indicated, much of the paralysis disappears, but a residue can always be found by a careful examination. The paresis or paralysis of the facial is almost always limited to the inferior distribution of the nerve. The superior extremity assumes peculiar attitudes. The forearm tends to bend on the arm, and the hand on the forearm, but these conditions or attitudes are only rarely spastic in character, as is observed in capsular lesions. This point seems an important diagnostic one to the writers who have observed cases presumably of lenticular lesion in which the non-spastic limbs assumed special attitudes, simulating those seen in spastic hemiplegia. The three portions of the upper extremity are usually about equally affected with the impairment of power. Gross oscillatory tremors may result from attempts to use the limb. In the lower limb, as in the upper, the paresis is usually about equally distributed in its three parts. It is rarely, if ever, a complete paralysis. The patient feels as if his paretic leg would give way; sometimes he can walk and sometimes not; when he walks his steps are slow and short. The leg of the paretic side is a little flexed on the thigh, the foot striking the

ground, without, however, performing the classic movement peculiar to capsular lesions.

Mingazzini lays stress upon the fact that the majority of his cases with the syndrome of hemiparesis could be referred to softening of the putamen, situated near, and in correspondence with, the anterior limb of the internal capsule and not with the posterior portion. The lesions being somewhat removed from the pyramidal tracts, therefore their effects when these lesions are hemorrhagic cysts cannot be attributed to pressure upon these tracts. In addition, Mingazzini points out that in most of his cases the lesions were foci of softening which did not exercise pressure on neighboring parts. A still further point in favor of the paralytic disorders being due to lenticular lesions and not to pressure exerted upon the capsule is the fact that these affections, even if they are not marked, are persistent and continue until death. Although partial paralyses, they are not transitory.

The motor function of the lenticula, says Mingazzini, deserves to be called supplementary if it is borne in mind that the motor disturbances in comparison to those produced by the destruction of the pyramidal region of the internal capsule are very mild. Mingazzini does not attempt to decide whether the motor fibers which originate in the lenticula join with those of the internal capsule; whether the neurones of which they are part originate in the motor cortex or in the lenticula itself; and whether if they originate in the lenticula they come in contact with neurones derived from the cortex. These are matters for future research to determine, especially by the studies of experimental and other degenerations. His claim is simply that the lenticula is a motor organ of some sort; that it possesses real motor function. He refers to Johansen's faradic excitation of the lenticula as confirming his own results and views.

Disturbances of speech, according to Mingazzini, are often associated with lesions of the lenticula. He says with some reserve that he believes the dysarthric disturbances dependent on lenticular lesions arise only when these lesions are situated on the left side. He has recorded cases in which the dysarthric disturbances were absent even when the lenticula was injured on both sides. His explanation of such observations is that the fibers destined for the movements concerned in speech run in circumscribed zones, as it is true that the fibers destined for the face, the

upper and lower limbs, run also in circumscribed zones; in other words, that the motor zone of the lenticle is subdivided as is the cortical motor zone.

Discussing in detail some of his observations regarding speech, Mingazzini says that disturbances of articulation under the form of elisions, initial pricking and bad pronunciation of the dentals were signalized in one case in which the external member of the globus pallidus on the left was destroyed; and in another case in which there was a softening of the left putamen. But it was impossible to find any disorder of speech in still another case in which there was a hemorrhagic focus on the external margin of the right putamen. Disturbances of articulation were found in a case in which there was a loss of substance which occupied the more external limit of the left putamen, on a level with the middle part of the colliculus caudatus; and these were not found in another case in which the hemorrhagic focus occupied the medial and inferior part of the globus pallidus of the left; and not in still another case in which the lesion was in the base of the putamen and in the middle part of the external capsule of the left. These results demonstrate again that the lesions of the right lenticular nucleus have no effect on speech, and that those of the left lenticular nucleus give rise to disturbances of articulation only if situated at fixed points.

VIEWS OF SCHÄFER, VON MONAKOW AND OTHERS.

Coming to one of the most recent physiological works, the views of Schäfer,¹² regarding the functions of the striatum (lenticle and caudatum) are stated in the following manner:

"Cajal describes axones of many of the pyramidal cells of the frontal lobe as passing to the corpus striatum. After destruction of the frontal lobe in dogs and monkeys, Marinesco found numerous degenerated fibers in the corpus striatum and especially in the caudate nucleus. On the other hand, the corpus striatum receives numerous fibers from the thalamus and the sub-thalamic region, and probably sends out centrifugal fibers downwards, along the motor tract; but these have not hitherto been satisfactorily traced; some appear to go to the substantia nigra. The

¹²"Text Book of Physiology," edited by E. A. Schäfer, LL.D., F.R.S., Vol. II., Edinburgh and London, 1900.

corpus striatum is generally believed to act as a center for the higher reflex movements, and to be in close association with the Rolandic area, but the experimental grounds for this belief are still lacking. Morphologically, the corpus striatum is regarded as a part of the cortex."

Von Monakow¹³ says that a lesion taking in almost the entire caudate nucleus, leaving the internal capsule free, or even the lenticular nucleus, does not of necessity cause a persistent hemiplegia. He continues that Reichel¹⁴ has shown that a symmetrical softening of both lenticular nuclei may occur without clinical manifestations. As a rule, hemiplegia with the involvement of the facial and hypoglossal occurs in extensive lesions of the caudate and lenticular nuclei. It is seldom complete and only persistent when the internal capsule, that is the premedial portion of it, is implicated. Some areas of softening, either in the lenticle or in the caudatum, usually are latent. Irritating foci, for example tubercles, cause occasionally early hemiplegic contraction. Aside from the hemiplegia,—though together with this in connection with lesion of the caudate and lenticular nuclei—occasionally occur hemianesthesia, tremor, forced laughter, choreiform movements, vasomotor disturbances (elevated temperature, reddening of the skin, etc.) and central pain in the opposite half of the body, especially in the arm. But all these phenomena, as a rapid glance through the literature on focal diseases shows, may occur without direct lesions of the ganglia of the forebrain. Occasionally they appear following lesions in the neighborhood of the posterior internal capsule, in the posterior portions of the thalamus, and in the tegmental region. Nothnagel, for instance, says von Monakow, attributes vasomotor disturbances accompanying hemiplegia to involvement of the posterior internal capsule. Numerous observations speak, however, for the closer relation between elevation of temperature in the paretic extremities of one half of the body and disease of the caudate nucleus in the opposite half brain. Experimental investigations also are favorable to this view. We know that irritation of the caudate nucleus in animals may be accompanied by pronounced, though transitory, elevation of temperature. Various investigators have observed similar elevations of temperature on the opposite side to the foci

¹³Von Monakow, C. "Gehirnpathologie," second edition, Wien, 1905.

¹⁴Reichel. Wiener Medicinischer Presse, 1898. "Diseases of the Caudate Nucleus and Lenticular Nucleus." Cited by von Monakow.

of disease in man. In such cases edema of the skin was also present in the extremities. In all these cases the lesion, even microscopically, was not limited strictly to the caudatum.

Choreiform movements, says von Monakow, were years ago associated occasionally by this or that investigator with disease processes in the caudate or the lenticular nucleus. Ellischer, Flechsig and others have seen in those who have been affected with chorea and have died, peculiar, strongly refractive, often mulberry-shaped, bodies in the lymph sheaths of the vessels in the portions of the brain mentioned. Wollenberg found similar formations in the lenticle of persons who had not suffered from chorea, and therefore disputes the causal relation between chorea and these bodies. Heholt observed tremor in the extremities in a case of lesion of the putamen. Such isolated observations, according to von Monakow, are of little value so long as they relate to the chance occurrence of focal disease. At all events, he says, a uniform relation has not yet been established between choreiform movements and disease processes limited to the caudate or lenticular nucleus.

Deroubaix,¹⁵ in a paper on spasmodic laughing and weeping, reports one case with necropsy which is of great interest in connection with the pathology of this emotive syndrome, and with the general discussion of lesions of the lenticle. The patient was a man thirty-eight years old who had had a stroke causing left hemiplegia, but without any aphasia or dysarthria. He exhibited spasmodic laughter followed by weeping and accompanied by associated movements in the paralyzed left arm. Death was from a second stroke. Necropsy showed a large area of softening which had destroyed the lenticular nucleus, the anterior limb of the internal capsule, and all the white matter of the frontal lobe and of the central region, to the cortex on the right side. The thalamus, genu and posterior limb of the internal capsule were not affected.

Some of the conclusions drawn by Deroubaix are as follows: That the fibers concerned with the function of emotive expression probably pass outside of the pyramidal fibers like those concerned with the function of coordination and tonus; that the thalamus is the seat of the automatic movements of emotive expression, es-

¹⁵Deroubaix, M. A. *Journal de Neurologie*, Vol. XI., No. 5, 1906.

pecially of laughing and crying; that laughing and weeping are under the control of the cortex; that the cortico-thalamic fibers pass in front of the knee of the internal capsule, probably by the lenticular nucleus. Deroubaix therefore did not attribute the spasmodic laughing and weeping in his case to destruction of centers in the lenticula, but of fibers passing from the cortex through the lenticula to the thalamus. He speaks especially of the facio-thalamic bundle.

With regard to this explanation it is not altogether acceptable, as Mingazzini believes he has shown and as facts supplied by others indicate, that the lenticula is a motor organ and has distributed through it zones and centers of particular kind for movements of various parts of the body, including the movements connected with the expression of emotion. This being the case, destruction of such centers, especially if on both sides, would give rise to such a lack of control of the movements of expression as to cause the involuntary or spasmodic laughing and weeping.

In one or two of our cases of lenticular lesion involuntary emotion was present.

In connection with the question of speech disorder from lenticular lesion it will be observed in the case of Deroubaix that the lesion was right sided.

Pagano,¹⁶ by injecting solutions of curare into the caudate nucleus has concluded that:

(1) Excitation of the anterior third and of the middle third of this nucleus provokes in dogs something very like the emotion of fear. This effect is best brought out when the injections reach the internal half of the organ. All the characteristics of this emotion are present, the gesticulations, the play of the physiognomy, the cardiac and respiratory phenomena, the actions of the intestines, and of the bladder, the state of the pupil, and the effect of threats and noises all go to form conclusive evidence of this emotion.

(2) The excitation of these points, but especially of the middle third, provokes a strong erection of the penis which appears immediately after the injection and persists until death.

(3) The excitation of the anterior extremity of the nucleus

¹⁶Pagano. *Riv dipatol. nerv. e. Ment.*, July, 1906. Reviewed by W. W. Ireland in the *Journ. of Mental Science*, January, 1907, p. 175.

produces an agitation which presents the appearance of fear modified with anger.

(4) The excitation of the posterior third of the nucleus provokes a series of manifestations of anger; the grinning and barking, the readiness to attack and bite, and the whole attitude leave no doubt as to the nature of the emotion.

(5) The excitation of the outer part of the anterior third of the nucleus, besides some emotional disturbance, provokes in a greater degree intestinal and vesical phenomena.

The nucleus caudatus according to these experiments is a center of some of the emotions.

Pagano has shown the existence of distinct motor centers in the cerebellum by the same method.

THE NUMBER AND CHARACTER OF THE CASES STUDIED BY THE WRITERS.

The cases which follow are a few out of a considerable number which have come under the observation of the writers, in which lesion of the lenticular nucleus was found at necropsy. In the majority of these cases the lesion, usually a hemorrhagic cyst or an old area of softening, has not been confined to the lenticula; indeed, in most of them several parts of known functional importance, and others whose functions are still in obscurity, were involved in the destructive area of disease. The reason of this is of course easy to recognize, and resides in the fact that the blood supply to the great basal ganglia is from branches of the middle cerebral artery, which also sends branches to various regions both of the cortex and of the interior of the brain. The writers have realized the same difficulty that has been realized by others in reaching conclusions regarding the functions of the striate bodies, because of the infrequency of lesions isolated to them. Even in the cases chosen to illustrate some of the points of this paper, the lesions present were scarcely in a single one absolutely limited to the lenticula. In two or three, however, this was so nearly the case that they have felt justified in drawing some inferences regarding lenticular function.

The points which the writers have attempted to elucidate or at least on which they have tried to obtain some light through the study of their cases, circle around the subject of the mechanism of cerebral speech and that of the representation of motility.

CASE I. Apoplectic Attack Causing Right Hemiplegia and Loss of Speech—Partial Recovery of Motility and Speech—Some Mental Slowness and Confusion—Dysarthria—Partial Word Dumbness—Word Deafness Present but only Slightly Marked—Hemorrhagic Cyst involving the entire Lenticula, the white matter of the Insula and the anterior part of the First Temporal Convolution, and slightly the Posterior Limb of the Internal Capsule.

M. S., aged forty-eight years, white, born in Ireland, was admitted to the men's nervous wards of the Philadelphia General Hospital August 21, 1903.

On admission the patient complained of headache and also of more or less weakness and helplessness in his right leg and at times in his right arm.

No family history and no history of important illness previous to that producing his condition on admission could be obtained.

The patient made a statement that he had had fits after an injury to his head, after which he had also developed some difficulty of speech.

He also said that three months ago he came home feeling in his usual health, and after supper became dizzy and fell on his face. He said that he did not know anything for two weeks, but when he recovered his mind, he found that he could not use his right leg or arm and could not speak at all. He remained unable to speak for about two months, since which time his power of speech had gradually returned, but he still spoke with difficulty.

The patient was a hearty, robust-looking man. His tongue was moist, slightly coated; pulse of good quality. His heart action was irregular. The lungs showed good expansion.

The pupils were equal, reacted well to light, but slowly to accommodation.

Examination made September 16, 1903, showed but little loss of power in the right arm or leg; the grip on the right side was, however, slightly impaired. Station was good and gait steady. The reflexes were normal. No note as to sensation was recorded.

On February 24, 1904, a note was made by one of the physicians in attendance which stated that in his last apoplectiform attack he had some spasm in the right arm and leg; and also that he had sensory aphasia; and although he did not miss any words he occasionally missed a syllable.

On February 25, 1904, the record stated that his gait was fairly good, but slightly stiff; that he dragged the right foot slightly. The knee-jerks were spoken of as "spastic" with slight attempts at clonus on both sides; station was good with the eyes open or closed; the pupils responded to light and accommodation; no palsies of ocular muscles were present; the tongue protruded to the right; the naso-labial fold was slightly deeper on the right

side. The grip was now said to be better on the right than on the left side.

The patient's mentality was noted as "slow"; he required more time to comprehend questions. He could not read nor write because of lack of education. He appreciated the purposes and uses of objects. He recognized a book and named it as "book." He called a gold watch a "dollar." When pointed to the time, which was 12.20, he said it was "after two"; he could not specify the minutes. He could say the days of the week slowly. When told to close his eyes he grasped his hand; when asked to point to a chair, he picked up his hat. He easily confused the name of one object with that of another. He recognized what spectacles were and said that he needed them himself; he could not, however, name them. He could not name

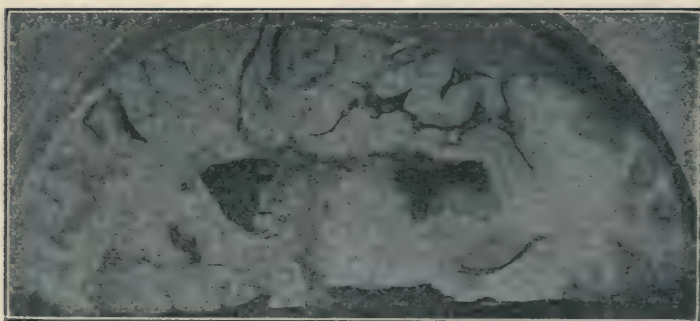


Fig. I. Cyst of the Left Lenticular Nucleus and External Capsule. Motor Aphasia Incomplete (Case I).

a lead pencil. He recognized a pen, could not name it at first but after a little effort was able to name it. He was also asked to name various other objects, as a key, a ring, and gloves, but failed to do so; but he named some objects of wearing apparel correctly.

Astereognosis was not present.

On December 21, 1905, he was found in a state of collapse and the following facts were noted: His eyes were turned upwards and oscillated slightly from right to left; the face was drawn slightly towards the left side. The right arm and leg dropped lifeless when raised. There was some spasticity in the right arm. The reflexes were exaggerated. On December 22, 1905, the patient was conscious again and seemed to understand all that was said to him, and a note said "were it not for his partial aphasia would be able to answer intelligently."

The heart sounds were very good, the second sound being exaggerated, especially the aortic. Pulse at the wrist was strong,

of rather high tension and moderately rapid. The patient did not respond to pin pricks. Upon catheterization only one half ounce of urine was obtained.

On December 23, 1905, the patient was in fairly good condition at 10 A.M. At 11.15 he was found rigid, with legs extended and arms held close to the body, the forearm being flexed on the arm. His face was markedly cyanosed. The jaw was rigid, respirations had ceased and the pulse could not be felt at the wrist. The heart sounds could not be heard. A minute or so later the patient relaxed and was dead.

A section made at the level of the upper part of the basal ganglia revealed a cyst in the left hemisphere, which had implicated the middle and posterior part of the left lenticular nucleus, and extended to, but did not involve, the cortex of the island of Reil. It extended ventrally to 0.5 cm. (1.5 in.) from the anterior limit of the lenticle. It extended to, or possibly slightly involved, the posterior limb of the internal capsule. At a level 0.5 cm. (1.5 in.) lower the cyst extended from its posterior end into the anterior part of the white matter of the first temporal convolution, but did not involve the cortex, nor the posterior half of the first temporal convolution.

The temporary right hemiplegia with residual very moderate hemiparesis was probably due to the lenticular lesion, although it is possible it may have depended on the slight involvement of the internal capsule. The temporary, somewhat complete, aphasia, was probably due to the effects of this lesion on neighboring parts, some of the motor defect in speech being, however, probably due to the lenticular lesion. The partial word dumbness could not have been due to lesion of Wernicke's zone, as only the anterior part of the first temporal convolution was involved. As the posterior part of this convolution is considerably higher than the anterior part, the fibers from the former could not have been greatly involved by the extension of the cyst at a low level into the anterior part of the first temporal convolution. It is more probable that some of the symptoms are to be attributed to injury of fibers in the external and extreme capsules which stand in relation to the posterior part of the first and second temporal convolutions.

CASE 2. Apoplectic Attack causing Right Hemiplegia and almost complete aphasia—Only a few Recurring Utterances retained—Marked Word Deafness at First—Probably some Word Blindness which soon Disappeared—Power of Humming Melodies retained—Continuous Improvement under Systematic Training—Spontaneous Language largely regained—Ability to write with unparalyzed arm acquired—Mentality practically unaffected—Death after four years from Thrombosis of Superior Mesenteric Artery—Dura thickened and adherent—

Cyst destructively involving the Insula, the Lenticula, the Anterior and Posterior Limbs of the Internal Capsule, the External part of the Thalamus, and the White Matter of the Anterior Half of the First Temporal Convolution at a little distance from the Cortex—The Area of Broca and Subjacent White Matter and Wernicke's Zone not involved.

This case is one, the clinical details of which were in part given by one of us, Dr. Mills,¹⁷ in a paper on the treatment of aphasia by training, published nearly two years before the death of the patient. A contribution giving some facts regarding his condition and attempts made at training him for his aphasia before he was seen by Dr. Mills has also been published by Mrs. Kate Herman.¹⁸

In the account which follows the writers have summarized the most important facts bearing upon his aphasia and hemiplegia from the papers just referred to, and also from communications received from the patient's former secretary, Miss Dorothy C. Falk, and his former professional and business associate, Dr. H. C. Hays. As this contribution is intended to deal more especially with the functions of the lenticula and the symptomatology resulting from a lesion of this ganglion, the writers have thought it better to concentrate attention upon the facts demonstrating his type of aphasia, rather than upon the efforts at re-education of his language and the results of these efforts. The case is unusually valuable because of the fulness of the history obtained from the different sources to which reference is made, and also because of the intelligence and education of the patient which allowed a more complete record of his speech defects.

The patient was a physician, forty-five years old, residing in one of the western states. In July, 1902, he had an attack of right hemiplegia with almost complete aphasia. He was first under the instruction of Mrs. Kate Herman from June 15, 1903, to July 15, 1903. At the beginning of this period his vocabulary consisted of only four expressions, namely "yes," "no," "but," and "O God." "Yes" and "no" were commonly used correctly. He sometimes added the word "but" to "yes" or "no" apparently when the "yes" or "no" did not seem to him to express his meaning, or in order to give some force to what he was trying to say. At other times he would use the expression "O God." He often used pantomime in an unmistakable manner. When he got possession of a few useful words and phrases he discontinued this expetive. Dr. Mills does not recall his

¹⁷Mills, Chas. K. "The Treatment of Aphasia by Training." *Journ. of the Amer. Med. Association*, Dec. 24, 1904.

¹⁸Herman, Mrs. Kate S. "Heilung der Aphasie durch den Artikulationsunterricht." "Eos." *Vierteljahrsschrift für die Erkenntniss und Behandlung jugendlicher Abnormer*, Heft 3, 1906.

having used it when he was under his care. His memory for persons, places and occurrences was good. He was often very insistent in his efforts to make himself understood. His instructor was soon convinced that his periods of concentration should be short and his exercises for improving his language varied. Word deafness was quite marked at this time, but was not complete. He seemed at times to be partially word blind, but this defect was probably comparatively slight if it really existed, as even early in the efforts to train him he was much helped by seeing the words which were used in training him either in script or printing. It was noted that he could assist himself in understanding what was read from a newspaper by tracing the words with a pencil as the paper was being read.

When he heard music which he knew, he would hum the melody and occasionally articulate some of the words of the song. One day when he was trying to relate an historical occurrence he could not pronounce the word "Maryland" intelligibly until it fortunately occurred to him to murmur the song, "Maryland, my Maryland."

His voice was not impaired,—a rather interesting fact in connection with the functions of the lenticula. He was trained systematically after the manner used with dumb children. The formations and combinations which he found most difficult were those with which such children struggle most, namely, *k*, *g*, and *ng*; *th* also gave trouble. He was made to repeat the names of objects in his environment, and his teacher states that the patient would succeed one day in using such words as *house*, *cow*, *tree*, *bird*, etc. and at another time would be unable to recall these, unless helped a little at the beginning. Gradually verbs were taken up and longer and longer sentences tried. With his first instructor, as later with Dr. Mills, it was found that he experienced his greatest difficulty with adjectives, pronouns and articles, and words generally which expressed no mental image.

He was taught to write with his left hand, making continuous and notable progress. He repeated aloud what his instructor wrote for him.

When he first consulted Dr. Mills in October, 1903, his right-sided paralysis, though still marked, was much improved. No loss of sensation and no affection of the bladder or bowels were present. The face was the seat of a moderate right-sided paresis, the tongue not deviating to either side. The deep and superficial reflexes were of the usual type found in cerebral hemiplegia—exaggerated knee-jerk with front tap and ankle clonus and the Babinski response being present on the right side.

When tested with Wyllie's physiologic alphabet, it was found that he could repeat the vowels *a*, *e*, *i*, *o* and *u* well, having been previously trained in their pronunciation, but he could not remember them from day to day. In like manner he could repeat

the consonants of the alphabet, but could not recall them spontaneously. He was partially word deaf, having regained word hearing to a considerable extent by time and training. He could understand most familiar expressions, but if a strange or unusual word were used he did not understand it. He was also apparently partially word blind, having relearned to read considerably before coming under observation. With his left hand he could write the words that he was able to read. He could not understand the meaning of such prepositions as *of*, *to*, and *for*, if pronounced or read. He could not spontaneously write these words when asked, but could copy them.

In connection with a study of his reacquisition of language, particular attention was paid to the degree in which he regained different parts of speech. It was noticeable that in his efforts to talk he chiefly used his nouns and verbs, occasionally employing a pronoun. The words which expressed the qualities of things, which join and relate things to each other and to actions,—in other words, adjectives, adverbs, conjunctions and prepositions,—were still almost entirely eliminated from his vocabulary. Auxiliary verbs like *to have*, and *to be*, in all their modes and tenses, were apparently without any meaning to him. The word *is* or *was* occurring in a sentence seemed to worry him. In brief, this patient was a hemiplegic aphasic, the motor aphasia having at first been almost complete; the sensory aphasia partial. An appreciable degree of recovery from his aphasia had occurred from the first under the method of training by repetition and by efforts at spelling, reading and writing.

Persistent efforts at training were made. The measures employed were the facial and other gymnastic methods used with the deaf and dumb, the method of repetition from dictation, of copying, the use of Wyllie's physiologic alphabet, of a phonetic reader, a language primer and grammar. An interesting point in the history of the attempts at this man's re-education in speech was the fact that he made much use of a small dictionary. He made considerable advance by looking up the definitions of words, and when he found words and expressions in the definition which he could not comprehend, these in their turn were looked up in the appropriate places.

After the patient's return home communications were received from time to time regarding him. He steadily improved, although at first his progress was comparatively slow. In a short time he was reading his newspapers and magazines and understanding fully what was in them. Six months after he left Dr. Mills word was received that he was acquiring more voluntary expressions every week. A noticeable feature now was that when he was told to say anything instead of repeating it verbatim, he would express the idea in his own words. He read much and played whist and other games, and had just finished transacting

some business for the institution, the first which he had done of any importance since his illness. A little later, in describing his improvement, the physician associated with him in his work, said that his use of words to express an idea was much more general than had been the case heretofore. He now expressed in his own words answers to questions in the form of a sentence rather than a single word, and it was noticeable that he was able to use a different word to express the same idea. At times he made sentences of considerable length, using nouns, verbs, adjectives, prepositions, etc. Thorough and systematic training with the assistance of his instructors and members of his family was kept up, the patient helping the work very much by his own determination and originality in methods. He continued to use his dictionary. His teaching was much along the line of both what is known as the word method and also that of spelling by using each letter as in the older method.

Many of the details of the methods used to improve his language, and of the time spent in these efforts will be better reserved for an additional paper. With regard to the re-education in language this case is the most interesting that has ever come under our observation. Nearly four years elapsed between the time of the apoplectic attack causing his hemiplegia and aphasia, and his death. During much of that time with interruptions sometimes extending over a few weeks, or even months, the earnest, formal retraining of his speech was carried on, and even when this systematic instruction was not resorted to, the patient, a man of much originality and energy, was always trying to re-educate himself in the use of language. His former secretary calculated that during the four years he was given about one and one half years of training. As a rule only one to two hours daily were given to formal instruction.

It is interesting to note in connection with the question of intellectual deficit in aphasics, which has become so prominent through the views of Marie, that this patient retained throughout his mental vigor and general capacity. His deficiencies seemed to be entirely those conditioned by his defects of speech, meaning by this, of course, his word deafness, his early slight or moderate word blindness, and his difficulty in correlating concrete concepts with each other as well as his very pronounced motor aphasia. About nine months after his attack the sanitarium, of which he was the proprietor and chief physician, burned down. Measures were at once taken to rebuild it, and as bearing on his mental condition, his secretary wrote Dr. Mills that he was always able to figure on any proposition just the same as before his illness but he could not speak it, all his figuring for his sanitarium being done in a very accurate and clear-sighted manner.

The improvement in his speech was continuous up to the time of his last illness.

He had no second apoplectic attack. He had, however, three distinct epileptiform seizures, at long intervals, during the last two or three years before his death. No increase either of his paralysis or of his aphasia occurred as the result of these attacks.

His general mental state was good. At times there were indications of moroseness, some irritability of temper and impatience when he was not immediately understood, and occasion-



Fig. 2. A case of marked motor and sensory aphasia. The area of Broca is intact and the subcortical white matter was not involved in the primary lesion (Case 2).

ally when much discouraged, he spoke of suicide. He showed no marked change in disposition which could be noted as altogether different from his former self. His general physical health remained good.

The conditions and symptoms preceding his death could in no way be attributed to the cerebral lesion causing his aphasia and hemiplegia, although the changes found in the aorta and the superior mesenteric artery might have some relation to the alterations in the vessels of the brain which were present at the time of the cerebral attack. He died about four years after the onset of his aphasia and hemiplegia, from thrombosis of the superior mesenteric artery, as determined at the post-mortem

examination. His symptoms were those of shock and great abdominal pain.

The necropsy was made by Dr. Philip Hillkowitz of Denver, Colorado, at which place the patient was at the time of his death. We are much indebted to Dr. Hillkowitz for the account of the gross findings. The body was well nourished, with considerable adipose tissue; muscular development good; the heart showed



Fig. 3. The lenticular nucleus and the island of Reil were destroyed. The sclerosis extended anteriorly to the white matter of Broca's area, and posteriorly to the posterior extremity of the island of Reil (Case 2).

slight thickening of the mitral valve; numerous small plaques were found on the aorta, above the valves and around the opening of the coronary arteries. Marked hyperemia was present around the opening of the right coronary artery. The lower lobe of the left lung was strongly adherent to the chest wall and diaphragm. The left kidney was very much puckered on its surface. A slight scar was found on the external border, about 1 cm.

(2.5 of an in.) in diameter, extending into the interior. The cortex was slightly diminished and was pale in color; the right kidney was similar to the left with the exception of the scar. The intestines showed a dark purple color extending all over both the large and small intestines. The superior mesenteric artery showed a thrombosis which could be followed in the branches of the vessel.

The dura was much thickened and adherent in many places to the membranes beneath. The membranes over the occipital lobe were largely congested. An elliptical cavity 5 x 2.5 cm. (2 x 1 in.) was found in the left cerebral hemisphere. The description of the parts destroyed by this lesion is given in the following paragraph after careful study of the specimen by Dr. Spiller, the brain having been forwarded, after hardening in formalin, to the Laboratory of Neuropathology of the University of Pennsylvania by Dr. Hillkowitz, who however had made an examination at the time of the necropsy. The description of the position and extent of the lesion differs in only one particular from that furnished by Dr. Hillkowitz, although this is one of some importance. Dr. Hillkowitz speaks of the lesion as involving a portion of the inferior frontal convolution (presumably meaning by this the third frontal). Although the cyst extended close to the third frontal it did not involve the cortex of this convolution.

A large cyst was found in the left cerebral hemisphere. In a horizontal section through the lateral ventricle and upper part of the basal ganglia, the sclerosis was found to extend to 1.5 cm. (3.5 in.) from the cortex of the convolution posterior to the ascending limb of the fissure of Sylvius. The area of Broca and subjacent white matter were intact, although fibers passing to or from the foot of this area were probably implicated. The cyst had destroyed the island of Reil, the whole of the lenticula, the anterior and posterior limbs of the internal capsule and the external part of the thalamus. The head of the caudate nucleus was very slightly affected. The gray matter of the first temporal convolution was not involved, although the white matter was involved at a little distance from the cortex.

The part of the first temporal convolution destroyed was the white matter of its anterior half. To be exact, the posterior border extended to the beginning of the innermost part of the middle portion of the first temporal convolution, in a section made through the upper part of the basal ganglia.

This case, although one of great value in connection with the discussion of the subject of aphasia, loses some of its importance as regards the functions and symptomatology of the lenticula, because of the fact that not only was this ganglion destroyed, but so many other functionally important regions. Any speech or paralytic disorder if dependent upon the lenticular

lesion, was probably covered by the effects of destruction of other parts. Broca's convolution was not involved, as was not the Rolandic operculum.

THE LENTICULAR ZONE OF MARIE AND MOTOR APHASIA.

Some light is thrown on several of the points of great interest in connection with the current discussion of the subject of aphasia, springing out of the somewhat revolutionary contributions of Marie, by the clinical and pathological facts observed in the case just described. Marie has laid much stress upon the lenticular zone. In defending himself against those who have charged him with giving too much importance to the lenticula in his theories of the pathogenesis of motor aphasia, he insists that the lesions producing the train of symptoms to which he has called attention are not due to isolated lesions of the lenticula, but to it and to those portions of its environment which he includes in this lenticular zone. It is important, therefore, to know just how he defines this zone.

In a horizontal section of the brain, he says, a transverse line drawn from the anterior angle of the insula to a corresponding point of the lateral ventricle will give its anterior border, while a line from the posterior angle of the insula to a corresponding point of the lateral ventricle will give its posterior limit. Thus he includes within this zone more than half of the thalamus, both limbs of the internal capsule, the greater part of the caudate nucleus (all except a part of the tail), the lenticular nucleus, and the island of Reil with the subjacent white matter. Marie does not attempt to distinguish between the symptoms of lesions of the striatum and of the white matter and cortex of the island of Reil. He defines his position concisely as follows:

A lesion implicating only the zone of Wernicke causes the aphasia of Wernicke:

A lesion of the lenticular zone causes anarthria, which is merely the pure motor aphasia of the writers, believed by them to result from a lesion of the white matter beneath Broca's area with the cortex intact. Articulate speech alone is affected, internal speech is preserved; the patient reads and writes and understands without difficulty spoken speech.

A lesion implicating the zone of Wernicke together with the lenticular zone causes the aphasia of Broca (great impairment

or loss of spontaneous speech, of reading and writing, and more or less impairment of the comprehension of spoken language).

The case just given fulfills these requirements for the aphasia of Broca. The patient was motor aphasic and agraphic in large measure and had great impairment of the power of reading, According to widely accepted opinion, the case should have exhibited also pure word dumbness and pure word deafness, because the lesion did not extend to the cortex either in Broca's area or Wernicke's zone.¹⁹

THE ROLE PLAYED IN THE PRODUCTION OF APHASIA BY LESIONS OF
DIFFERENT PARTS OF THE FIRST TEMPORAL CONVOLUTION.

In the discussion of the effects of lesions of the lenticular zone and the zone of Wernicke* in the production of aphasia, the importance of each portion of these zones must be carefully considered. It is only in this way that we can arrive at reliable conclusions on the mooted questions of the localization of the representation of the different elements of cerebral speech. The first and second temporal convolutions necessarily hold a prominent place in this discussion. It has been usually held that the higher auditory area, the so-called center for word hearing, is situated in the posterior portion of the left first temporal convolution, or of the first and second temporal convolutions, the focus of representation being about opposite the posterior upward turning of the horizontal branch of the Sylvian fissure. As a rule more stress has been laid upon lesions of the cortex than of the sub-cortex in the causation of word deafness, although of course the pure word deafness of Lichtheim and Dejerine has been attributable to injury of subcortical tracts proceeding to the cortical center from the basal centers and the auditory periphery.

In the two cases, the histories of which have just been given,

¹⁹Marie, Pierre. *Revue de Philosophie*, 1907.

*It might be well to give here Marie's definition of this zone. According to him it consists of the supramarginal and angular gyri and the feet of the first two temporal convolutions. Its limitations cannot be accurately determined, and it is impossible to say whether the symptoms accompanying a lesion of this zone are caused by the alteration of the cortex or the subjacent white matter; Marie inclines toward the importance of the white matter.

word deafness was undoubtedly present and somewhat persistent, although not permanent. In both of these cases the zone of Wernicke, as described by Marie, was not involved in the destructive lesion, this being confined to the white matter of the anterior portion of the first temporal convolution.

It is not necessary to call into service the theory of diaschisis of von Monakow to explain the word deafness in these cases. When word deafness is a symptom, the subcortex of the anterior portion of the first temporal convolution being diseased, while the cortex and subcortex of the posterior halves of the first and second temporal convolutions escape, the aphasic phenomenon finds its explanation in destruction of the paths of transmission to and through the external and extreme capsules to the insula and the center of Broca. One of the writers, Dr. Mills,²⁰ has reported a case, now well known in the literature of the subject, which shows that word deafness may result from a lesion limited to the posterior portion of the first or of the first and second temporal convolutions, chiefly to their cortex, and reference will next be made to two cases previously recorded by Dr. Spiller.²¹

Case 3. *Fracture of the Skull in the Temporal Region—Contusion and Disintegration of the Inframarginal and Supramarginal Regions—Hemiplegia, Hemianesthesia and Aphasia Nearly Total at First—The Disappearance of Paralysis and Improvement in Motor Aphasia, Marked Word Deafness and Paraphasia Persistent—Operation with Improvement—Relapse—Auditory and Visual Hallucinations—Persecutory Delusions—Necropsy Showing Destructive Lesion Chiefly Involving the Zone of Wernicke (the Supramarginal and the Posterior Portions of the First and Second Temporal Convolutions).*

The patient, a man twenty-nine years old, had suffered from a fracture of the skull which left him at first completely hemiplegic and hemianesthetic on the right side and almost totally aphasic. He recovered largely from the paralysis and to some extent from the speech disturbance, but word deafness and paraphasia were pronounced. He was probably word blind.

An operation revealed a stellate fracture with contusion and

²⁰Mills, Chas. K. "Lesions of the Superior Temporal Convolutions Accurately Locating the Auditory Center." *University Medical Magazine*, Vol. 4, November, 1891.

²¹Spiller, William G. "Lesions of the Left First Temporal Convolution in Relation to Sensory Aphasia." *Review of Neurology and Psychiatry*, May, 1906.

disintegration of brain substance, probably of the first and second temporal convolutions. Word deafness persisted, and later the patient became insane with auditory and visual hallucinations and delusions of persecution. The patient died in the insane department of the hospital and necropsy showed an area of sclerosis involving the posterior part of the first and second left temporal convolutions, and also part of the parietal lobe. It extended to the posterior part of the insula and to the optic radiations.

The necropsy showed the reverse picture of the lesions in cases 1 and 2 so far as the temporal lobe was concerned, the sub-

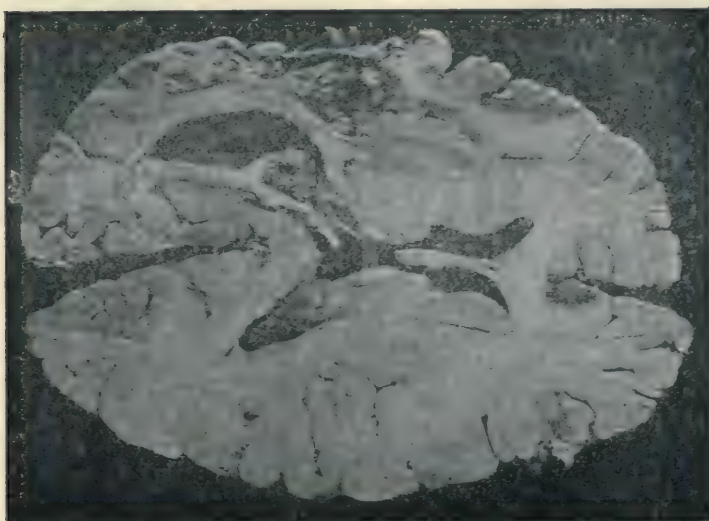


Fig. 4. Complete destruction of the first temporal convolution, with marked sensory aphasia. The lesion extends to the lenticular zone. (Case 3.)

cortex of the anterior halves of the first temporal convolutions being destroyed in these cases. Word deafness was pronounced but not complete. It would seem, therefore, from these three cases that either a destruction of fibers passing from the posterior part of the left first temporal convolution by way of the external capsule, or of these fibers plus a lesion of the cortex of the posterior part of the left first temporal convolution may cause word deafness. In the first two of these cases the word deafness was less persistent. It is to be noted that the sclerosis in case 3 involved the upper part of the middle portion of the left second temporal convolution.

Case 4. *Apoplectic Attack twenty-three years before death causing Hemiparesis and Disorder of Speech—A Second Apo-*

plectic Seizure fifteen years later—Slight Paraphasia and Paragraphia (Occasional Omission or Misuse of Words in Talking or Writing)—Word Deafness not present for several years before his death—No Facts as to Word Hearing at the Time and immediately after his aploplectic attack—No Word Blindness or Letter Blindness—No Mental Impairment except some Defect of Memory—Was Ataxic and had other symptoms of



Fig. 5. Complete and long-standing destruction of the left first temporal convolution without marked persistent sensory aphasia. The destruction extends to Marie's lenticular zone. (Case 4.)

Tabes—The Microscopic Findings of Tabes—Destruction of the Entire Left First Temporal Convolution except in the Angular Region—Destruction of the Insula except at its anterior pole—No Involvement of the Lenticula.

A man, about sixty-one years of age at the time of his death, twenty-three years before this time had an apoplectic attack. He was left with some disorders of speech and impairment of power in the right half of the body. About eight years before his death he had a second seizure. He became partly aphasic, could not write without making mistakes, walked with difficulty, and at times had difficulty in swallowing and in urination. He also had spells of hiccough and vomiting. He had cramps or twitchings in the right leg.

The patient, at least in a late period in the history of his case, had no difficulty in understanding words. He was sometimes at a loss for a word, although he could talk with comparative ease. He was liable to misuse words in writing, sometimes misspelling, but these defects were but little marked. He was sound mentally, except that his memory had failed somewhat. He was right-handed. He read constantly, evidently not being word or letter blind. He was ataxic and had other symptoms of *tabes*.

The left first temporal convolution was destroyed in its entire length except at the angular and supramarginal gyri. The sclerosis extended to the posterior horn of the lateral ventricle. It destroyed the cortex of the island of Reil and the white matter immediately subjacent except in the extreme anterior end of the island. It did not involve the lenticular nucleus. The lesion was very similar to that in the brain of the patient in case 3, except that it extended a little more inwards and considerably further forward, but left the supramarginal and left second temporal convolution intact. It did not cause persistent word deafness, at least the patient was not word deaf at the time his case was studied. About his condition as to word hearing at the time of his attack and in the earlier years after it, no record was made.

In the original report of this case, one of us, Dr. Spiller, has suggested that the only explanations for the preservation of word hearing in this case are that the center for this function was largely in the posterior part of the left second temporal convolution, or that the right first temporal convolution had been unusually well developed during the patient's entire life, and was capable of assuming the function of the destroyed left first temporal convolution. It seems improbable that the small portion of the angular and supramarginal gyri preserved could explain the retention of word hearing.

The preservation of mentality in this case, as in case 2, is of interest in connection with Marie's views as to intellectual deficit caused by lesion of Wernicke's zone. The history of the case emphasizes the fact, to which attention has been directed by one of us²² in a recent paper, that the degree of intellectual deficit in cases of lesion of this zone is largely conditioned by the intelligence and general capacity of the patient before the attack causing the aphasia.

THE INSULA AS A DISTINCT AND IMPORTANT FUNCTIONAL AREA.

One of the results of the acceptance of the views of Marie is to underrate the importance of the insula as a functional area or as a portion of a functional area concerned with speech. The

²²Mills, Chas. K. *THE JOURN. OF NERV. AND MENT. DIS.*, Vol. 34, July, 1907.

effort should in the first place be made to distinguish between the functions of the insula and those of the lenticula.

Some of the contributors to our knowledge of aphasia, and especially some of the earlier writers of the period beginning with Broca's discovery in 1861, have gone so far as to regard the insula as the sole cortical motor center for speech, a view which must fix our attention in connection with some of the findings given in this paper, although we are inclined to the opinion that it is only a part, but it may be a large part, of this cortical motor center. The numerous cases of lesion of Broca's convolution with motor aphasia, even if the lesion in many of these cases was not confined to this convolution, cannot be set aside. In one of our own cases (case 5) the lesion causing the motor aphasia was nearly equally distributed between the insula and Broca's convolution, Wernicke's zone, the lenticula and the capsules not being involved.

For the literature of the insula in its relations to speech we must refer our readers to such well-known works as those of Bateman, Wyllie, Dejerine, and others.

Dr. E. A. Spitzka²³ has contributed something of interest in this connection from the standpoint of the anatomist and morphologist. He has directed attention to the redundancy of the pre-insula in the brains of highly educated men,—men with great capacity for language both as a means of verbal expression and of reasoning.

The island of Reil is embryologically very different from the striatum. The latter rises from the base of the brain and is separated by a fissure, as Edinger says,²⁴ from the mantle or pallium, whose wall becomes thickened relatively late. The striatum develops in all vertebrates, but it is only in the higher that the pallium has a development of importance. The pallium later contains the cortex, on which depends all mental processes. In a figure of a frontal section through the brain of a human embryo at the age of two and one-half months, given by Edinger, the striatum rises free from the base of the brain, distinctly separated by a fissure from the external wall of the lateral ventricle. At a later embryological period (about the fourth month)

²³Spitzka, E. A. *Amer. Anthropologist*, N. S., Vol. 5, October-December, 1903.

²⁴Edinger, L. "Vorlesungen über den Bau der nervösen Centralorgane."

it is divided into the caudate and lenticular nuclei by the fibers of the internal capsule growing through it. The caudate nucleus projects free into the lateral ventricle, as does the lenticular nucleus also; but in later embryonal life the small fissure between the latter and the wall of the cerebral hemisphere becomes so narrowed as to be invisible; but even in adults the wall of the hemisphere can be separated from the outer part of the lenticular nucleus without tearing of fibers. This region of the early fissure is sometimes of importance in the fully developed brain. Hemorrhage occurs here very readily, and the blood, if not too great in quantity, fills the space between the wall of the cerebral hemisphere and the outer division of the lenticular nucleus. Edinger believes that the striatum has some of its fibers in the anterior limb of the internal capsule.

These remarks of Edinger explain why the external capsule is so often the area occupied by a hemorrhage or a cyst. In one of our cases (case 6) a cyst was found in each external capsule which was a *locus minoris resistentiæ*.

In another case studied by us,—not included formally in the cases given in this paper,—in which the lenticula was in an early stage of red softening, no separation of the lenticula from the island of Reil was seen when the brain was first opened, but during the process of hardening the putamen contracted from the white matter of the island, leaving a fissure in the external capsule.

The blood supply of the lenticula and of the insula and its subcortex seems to be distinct. We have preparations from a case (case 7) in which occlusion of the middle cerebral artery occurred after the supply to the basal ganglia had been given off. The point of thrombus formation is clearly demonstrated. The softening implicates the island of Reil and subjacent white matter, but leaves the lenticula intact. The patient could not speak during three weeks, but seemed to understand.

From these facts we may conclude that the function of the island and its white matter is distinct from that of the lenticula, and cases in which both structures are implicated may be wrongly interpreted.

Case 5. *Apoplectic Seizure causing Aphasia without Paralysis of Face or Limbs—Aphasia of Motor Type—A Few Recurring Utterances—Marked Paralexia—Some Retention of*

Writing—No Impairment of Articulation or Vocalization—An Old Cyst Involving only the Cortex and Subcortex of the Caudal Portion of the Left Third Frontal Convolution and the Insula.

Although the following case has been used by one of us, Dr. Mills,²⁵ in a previous article, and has also been referred to by Gordinier,²⁶ it has so much value on the negative side of the discussion of the functions of the lenticula, as this ganglion was not involved in the lesion, and also so much on the positive side of the consideration of the functions of the insula and Broca's convolution, both of which were destructively implicated, that we have thought it important to reproduce it in connection with our present series of cases. There is scarcely to be found in the literature of aphasia a more instructive case. Neither Wernicke's zone, the lenticula, nor the internal capsule was involved, and yet the patient showed motor aphasia of typical form with the retention of some power of writing and with the retention (or reacquirement, as the case was not seen at an early period after the apoplectic attack) of the power of reading silently.

The third frontal gyre and the insula were neatly involved in the same circumscribed lesion, the second frontal convolution, the internal capsule, the basal ganglia, and all other portions of the brain not being the seat of any old lesion.

The following citation and summary are taken from the paper in the *American Journal of the Medical Sciences*:

"This patient, after an apoplectic seizure, developed aphasia, apparently without preceding paralysis or any other symptoms of focal lesion like visual blindness and hemianopsia. When examined about nine years after the onset of the aphasia, he had almost complete inability to name persons and objects which he was able to recognize through all his special senses. He had also a marked form of paralexia. When he attempted to read, although he understood what he was reading, he repeated an absurd formula of a few phrases. He had limited spontaneous speech, even using short sentences without concrete nouns correctly. He could write many single words correctly, sometimes misspelling, however. He held his pen or pencil correctly and wrote with ease and firmness."

Many examinations of this patient were made. The case was clearly one of aphasia with marked verbal amnesia and general disorganization of language on its dynamic side. Grammatical expressions and combinations were lost. The patient could speak only in single words, or in short phrases, or very short sentences. What he said, however, he articulated and enunciated clearly.

²⁵Mills, Chas. K. *Amer. Journ. of Medical Sciences*, September, 1904.

²⁶Gordinier, C. H. *Idem*, 1903.

He had no paresis of the organs of articulation, enunciation or vocalization.

The only lesion to be seen in the lateral aspect of the left hemisphere was confined to the hinder part of the third frontal convolution. The lesion was probably part of an old hemorrhagic cyst. When the brain was sectioned at the level of the insula the insula was found to be largely destroyed by a lesion which was continuous with that in the caudal part of the left subfrontal gyre. Other parts were not involved, as shown by



Fig. 6. Case of motor aphasia without hemiplegia. The primary lesion involves the left third frontal convolution and the insula. (Case 5.)

this section and also a subsequent one made a little lower than the first.

It is worth while to refer in this connection to one of the most recent and valuable contributions on the pathology of aphasia—a paper which forms a part of the general discussion which has arisen since the promulgation of the views of Marie. In this paper, in addition to many important considerations regarding

focal lesions and the pathology of aphasia, Dejerine²⁷ gives the details of two previously unrecorded cases studied both macroscopically and in microscopical serial sections. In these cases the primitive lesions causing the aphasia were confined to the cortex and subcortex of Broca's convolution and to the second frontal convolution with a slight involvement of the insula and extreme capsule in one case. The zone of Wernicke, the lenticula and the capsules, except as mentioned concerning the extreme capsule, were not implicated if we exclude processes of secondary degeneration.*

The first of the two cases reported by Dejerine in this paper was one of motor aphasia, at first very marked, the patient retaining only "yes" and "no" and interjections. Any evidences of sensorial aphasia were very temporary. No paralysis, orolingual, facial or of the limbs, was present. The patient was motor aphasic without being dysarthric. Writing could not be tested, as the patient had lost his right arm by amputation and had not learned to write with his left hand. As time progressed, and before his death two years after the apoplectic attack, he had regained largely the power of speech.

The necropsy showed a lesion similar in some respects in its limitations to case 5 of our series, the insula, however, not being nearly so much involved. In Dejerine's first case the lesion destructively involved about two-thirds of Broca's convolution and to a very moderate degree the anterior portion of the insula and the related segment of the extreme capsule. The caudate and lenticular nuclei were not attacked by the primary lesion, as were not the Rolandic operculum and the external and internal capsules. Serial microscopic sections showed some involvement of the second frontal and of the corona radiata, with a considerable portion of the frontal lobe anterior to the precentral convolution. Degeneration was distinctly marked in the knee of the callosum. The temporal lobe was nowhere diseased. Softening was observed in the first and second frontal convolutions of the right hemisphere. The degeneration involved the anterior segment of the internal capsule, the fibers of passage through the striatum, and the occipito-frontal bundle. Degeneration of the anterior

²⁷Dejerine. J. *L'Aphasie Motrice et sa Localisation Corticale*. *L'Encéphale*, No. 5, May, 1907.

*We understand by Broca's area, which has been described differently by writers on aphasia, that corticosubcortical area which surrounds the ascending branch of the Sylvian fissure, including its bifurcations. This makes Broca's convolution include what Dejerine calls the orbital portion, the cap and the foot of the convolution. The foot of this convolution is the part situated between the precentral convolution and the horizontal and ascending branch of the Sylvian fissure.

fibers of the extreme and external capsules was also present.

In the second case of Dejerine, the patient, a woman seventy years old, had a left hemiplegia and some years later an attack causing motor aphasia. This motor aphasia as described was typical and persisted. The patient retained only a few expressions, such as "yes," "no," and "good day." She was not word deaf, word blind or letter blind, or if word blind at all only to a limited extent. She understood all that was said to her, even when she was spoken to rapidly. She could read what was placed before her, but had difficulty if the sentences were long or complex. She could not write, except her name, although she held the pen correctly. She could, however, copy and transfer print into script.

We shall omit the lesions found in the right hemisphere in this case, except to say that they were such as to account for her left hemiplegia and perhaps some other phenomena. In the left hemisphere a yellow plaque was found, so situated as to destructively involve the cap of Broca's convolution, and also in part the second frontal convolution. Serial microscopical sections showed that this primitive lesion did not involve the insula, the external or internal capsules or the basal ganglia. The white matter of the third and second frontal convolutions was considerably diseased. Degenerations were present much as in the first case, involving, for example, the anterior segment of the internal capsule, the radiate fibers to the thalamus and the knee of the callosum. The zone of Wernicke and all other parts of the left hemisphere were intact.

This case clearly shows that Broca's aphasia (motor aphasia of persistent form) may be due to a lesion involving the third and second frontal convolutions and the subjacent white matter in the primitive lesion, all other parts escaping.

The first of the two cases is of similar import, although the insula and extreme capsule were to a slight extent included in the lesion, and the patient recovered largely from his motor aphasia. This recovery may have been due to the restoration of the parts of Broca's convolution and the insula not included in the lesion, or as Dejerine suggests, the fact that the patient had lost his right upper extremity some twelve years before his death may have had something to do with the unusual facility with which the convolutional areas of the right hemisphere, homologous to those of Broca in the left, assumed the representation of motor speech.

Case 6. Right Hemiplegia with Exaggerated Reflexes on the Paralyzed Side—Attacks of Vertigo—Linear Cyst in the External Capsule of Each Hemisphere—Area of Sclerosis in the Middle of the Foot of the Left Cerebral Peduncle—Right Pyramidal Tract in Cervical Region Degenerated.

The notes of the following case were meager.

E. D., aged fifty-seven years, white, German, was admitted to the Nervous Wards of the Philadelphia General Hospital November 14, 1902. He had been a strong man and had worked as a sailor.

When admitted to the hospital he stated that he was paralyzed on the right side six years ago. He said that he had had attacks of vertigo every day for the last six months, in some of which he fell to the floor. The eyes reacted normally to light and in accommodation. On the right (paralyzed) side knee-jerk was excessive, slight on the well side. No ankle clonus could be obtained, but a slight Babinski response was present on each side.

The next notes in this case were not made until September

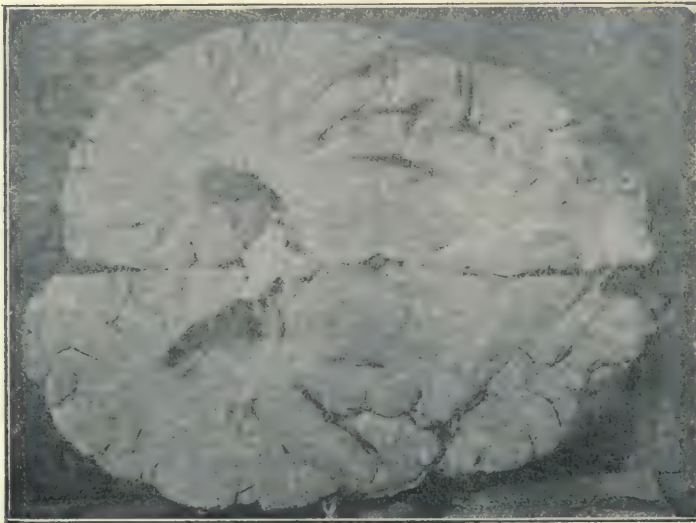


Fig. 7. A symmetrical cyst is present in each external capsule in the region of least resistance. (Case 6.)

11, 1903, when the records state that the patient was in a stuporous condition, unable to answer any questions and he did not seem to understand anything that was said to him. He had great difficulty in swallowing.

On September 15, 1903, the notes stated: He became weaker this evening and was no longer able to be out of bed. Incontinence of urine and feces was present. He died September 20th. The fresh spinal cord seemed to show sclerosis of both right and left pyramidal tracts. The brain showed some areas of softening of the right caudatum.

A horizontal section through the cerebral hemispheres nearly

at the upper border of the lateral ventricles showed a small hemorrhagic scar in the right hemisphere probably cutting some of the fibers that form the knee of the right internal capsule. This may have developed shortly before death. A linear cyst was found in the external capsule of each hemisphere in a section made through the upper part of the thalamus and lenticula. The cyst on the right side to the naked eye measured 2.5 cm. in length and 0.5 cm. in its widest portion (1 in. in length by 1-5 in. in width). It did not extend into the cortex of the island of Reil and encroached very slightly on the lenticula. Its anterior end was 0.5 cm. (1-5 in.) behind the anterior end of the lenticula. Its posterior end reached very nearly to the posterior end of the lenticula. The cyst on the left side was smaller than that on the right side. It was in exactly a corresponding position except that its anterior end was 1.5 cm. (3-5 in.) behind the anterior end of the lenticula. Its posterior end extended to the posterior end of the lenticula. It also encroached very little on the lenticula. Neither cyst extended into the capsule, and neither extended downwards as far as the level in which the cerebral peduncles appeared distinctly formed, nor did they change their relative positions. No cause was detected for the right hemiplegia.

Sections of the cerebral peduncles and of the cervical region of the spinal cord made by Dr. T. H. Weisenburg showed a small area of sclerosis in the middle of the foot of the left cerebral peduncle. The right crossed pyramidal tract of the cervical region of the cord was degenerated. The lesion producing this degeneration has not been found, unless it was the cyst in the external capsule, which was doubtful.

The history in this case is very meager, but we have retained it in this series because the notes record that the patient stated that he had been paralyzed in the right side six years previously, and made other assertions. He therefore probably was not motor aphasic, though we could not assert that his speech was normal. An old lesion, a cyst, was found confined to each external capsule and claustrum, and yet persistent or complete aphasia seems to have been wanting. The right hemiplegia might be attributed to this cyst in the left external capsule, but inasmuch as a small area of degeneration was found in the middle of the foot of the left cerebral peduncle, it seems possible that some other lesion in the left cerebral hemisphere has been overlooked. The lenticula on each side, to the naked eye, seems to have escaped.

This case, after all, is of chief importance as illustrating the points made when discussing the views of Edinger and others as to the embryonal, and even later, separations of the insula and its subcortex from the basal ganglia, the inference being that they are functionally, anatomically and morphologically distinct.

Case 7. Right Hemiplegia with Aphasia—Deep Reflexes but

little changed—Some Hypesthesia—Stereognosis not studied—Aphasia of Motor Type—No Word Deafness—Thrombus of Left Middle Cerebral Artery just above the place where the Vessels to the Basal Ganglia are given off—Softening involving the Cortex and Subcortex of the Insula, the Foot of the Pre-central and Postcentral Convolutions, the Entire Parietal Lobe, the Fibers to the Lower Part of the Callosum, the Optic Radiations to a small extent—The Lenticula and the Temporal Lobe were not implicated in the Lesion.

A. H., sixty years old, colored, was admitted to the Philadelphia General Hospital December 15, 1904, and died December 29, 1904. The only history that was obtainable was that she had not spoken for five days and since that time had been paralyzed in the right half of the body.

On examination the patient was found lying in bed with her eyes closed, making no attempt to move or talk. The face was drawn to the left over one-half of its extent. There was marked over-action of the occipito-frontalis of the left, while on the right movement of this muscle was very slight.

The eyes were directed anteriorly without deviation; the right pupil was contracted, and apparently immobile. She could not be made to accommodate; the left pupil was fixed; the media of both eyes were very clouded, especially on the left, which seemed to show a cataract. Arcus senilis was marked. She could not be made to move her eyes in any direction when told to do so, although she evidently understood the command, as she made an effort to obey by a slight turning of the head.

The tongue could not be protruded. She opened her mouth when asked to put out her tongue, which was pushed to the left and was slightly tremulous and somewhat coated.

The right arm was apparently completely paralyzed and fell helplessly when raised. The response was very feeble on tapping the triceps and biceps tendons. The right leg, which was paralyzed, gave a slight knee-jerk but no ankle clonus, although there was a suggestion of dorsal flexion of the great toe on plantar stimulation.

She slightly moved her paralyzed foot and leg occasionally when stuck by a pin. Attempts to elicit ankle clonus occasionally resulted in a flexion of the thigh on the pelvis. The left leg gave a very slight knee-jerk. No ankle clonus and no Babinski response were determined. This limb was moved somewhat more when stuck by a pin than was its fellow. There was no positive result on either side to stimulation by the Oppenheim method.

Under date of December 18, 1904, the clinical notes stated that there was no change in the patient's condition. She understood what was said to her, but could not answer. Her bladder and bowels were incontinent. Her pulse was very irregular and she

slept most of the time. She was mentally dull. Temperature at this date was still down, but respiration was up to 50; pulse was at 150. The right chest gave bronchial breathing and great impairment about its middle in the midscapular line. Both sides were full of large bubbling and smaller sized rales. She could be aroused by vigorous questioning, but could not answer, although she tried. She was very weak. She died December 29, 1904.

At the necropsy the following pathological conditions were found in the various organs:

Congestion of the lungs; chronic interstitial myocarditis; chronic congestion and interstitial splenitis; chronic congestion and interstitial nephritis with infarction and hemorrhagic and healed infarct of the kidney; chronic congestion and fatty fibrosis of the liver and general arteriosclerosis. The brain and spinal cord were removed for hardening and further investigation.

Later this investigation showed a thrombus occluding the left middle cerebral artery just above where the arteries to the basal ganglia are given off, so that the latter arterial supply was intact. A large area of softening was found which implicated the cortex and white matter of the island of Reil, leaving a narrow margin of white matter intact on the lateral aspect of the lenticula; the latter ganglion was not affected. The foot of the precentral and postcentral convolutions were softened, but the extreme anterior part of the island of Reil escaped. The softening extended very close to and probably slightly involved the optic radiations.

At a higher level and just above the lenticula the softening extended inwards so as to involve slightly the fibers going to form the lower part of the callosum.

The precentral convolution was softened only in its lower part, but the postcentral was more involved together with almost the entire parietal lobe. The temporal lobe was intact.

This case therefore showed very clearly that the arterial supply of the island of Reil is distinct from that of the lenticular nucleus, and that the lenticule may escape although aphasia is pronounced.

Cast 8. Apoplectic Attack causing Right Hemiplegia and Loss of Speech—Tongue protruded to the right—Right Hemianesthesia—Memory poor—Partial Recovery of Speech—Aphasia Motor in Type—Reflexes exaggerated on the paralyzed side—Linear Hemorrhagic Scar in the Left External Capsule—Lesion Involving also the Insula and Anterior Limb of Internal Capsule, the Lenticula and Posterior Limb of the Internal Capsule.

E. J., fifty years old, colored, born in Virginia, an expressman, was admitted to the nervous ward of the Philadelphia General Hospital in the service of Dr. Charles S. Potts, on June 12, 1903.

No family history or previous history of importance was obtainable. The patient denied having had syphilis.

Several weeks before admission he fell unconscious in his wagon and remained unconscious for three days. It was then found that the entire right half of his body was paralyzed. At first he was unable to make his wants known by talking, but speech gradually improved. His wife stated that for several weeks before the paralytic attack he had complained of headache.

On June 14th, two days after admission, it was noted that he could not move his right arm and leg, that his speech was slow and his memory apparently poor. In general appearance the patient was lean and weak. The pupils were equal; the light reaction was normal, the accommodation reaction poor. The pulse was intermittent, the chest normal, the abdomen tympanitic.

Under date of June 17th notes were made that he was anesthetic on the entire right side, the anesthesia extending about one inch beyond the median line on the left side. The conjunctiva on the right side was anesthetic. The patient was so stupid that it was difficult to get from him satisfactory answers. He could not see a quarter of a dollar when it was brought toward him from the right, but he recognized it at once when brought from the left, or when it was held in front of either eye. Dr. Howard F. Hansell, one of the ophthalmologists of the hospital, however, reported no hemianopsia. On the right side he could recognize cold, but invariably called *hot* "cold." He could not distinguish any difference between two test tubes containing hot and cold water when they were placed on the right side, but could at once when they were placed on the left side. He complained of much pain when the right arm or leg was moved, but these limbs could be pressed or touched with impunity and he did not complain of pain in these parts while they were at rest.

He had use of all parts of the left arm, although power in it was somewhat impaired. He could also move the left leg properly, but with some diminution of power.

Further examination showed absence of sensation in the right side to above the costal margin, except on stroking with a sharp point. Sensation was also absent in the right arm and upper part of the body, except on very deep sharp pressure. Sensation was present on the left side. The knee-jerk was increased on the right side and still more increased and spastic on the left. The biceps jerk was increased on each side. The tongue, which was tremulous, was protruded slowly and to the right.

On June 22d examination showed that hemianopsia was absent. The Babinski reflex was present on the right; the plantar reflex was present on the left. No ankle clonus was present on either side. The tongue protruded decidedly to the right, with tremor. The naso-labial fold was lessened on the right; the

mouth was very slightly drawn to the left. The pupils were equal, contracted to light and very slightly to accommodation.

Arcus senilis was present. The arteries were thickened and tortuous, the right radial being somewhat beaded. The pulse was intermittent, its tension moderate. The heart was irregular in action, the second sound being accentuated, the first sound clear but not proportionately loud. Considerable atrophy and contracture were present in the right hand; the patient complained of pain in moving his right leg. He had frequent micturition and had control of bladder and rectum.

He apparently understood all that was said to him, but was not always able to find words to answer. When the proper answer was repeated to him, he recognized it. There was therefore apparently some motor aphasia.

On July 15th he called the hot test tube "cold" on both sides, and still had complete absence of power in the right arm and leg.

On July 25th it was noted that he had made some improvement during the preceding five days, but on this afternoon he became very suddenly prostrate, dyspnea became marked, and the pulse weak and rapid. He died during this day at a time not mentioned in the clinical record.

Necropsy showed the dura and pia normal. The upper aspect of the right cerebellar lobe was edematous, grayish yellow in color with distinct loss of substance. Inspection of the ventricles was negative. The vessels at the base of the brain were sclerotic. The specimen was removed for hardening and further examination.

Other pathological conditions present were: Fatty degeneration of the liver and myocardium, thrombosis of the pulmonary arteries, chronic pleuritis, edema and congestion of the lungs, chronic nephritis, slight prostatic enlargement, fatty degeneration of the kidneys, especially of the right, deformity of the right foot, and ulcer on the right ankle.

A horizontal section made through the cerebral hemisphere at the level of the upper part of the basal ganglia showed a linear hemorrhagic scar in the left hemisphere in the external capsule. It extended forward as far as the anterior limit of the island of Reil but not beyond, its forks sending off a branch cutting the anterior limb of the internal capsule. It implicated the greater part of the left lenticula, and extended very near to the posterior limb of the internal capsule, although apparently it did not implicate this limb at this level. The cortex of the island of Reil was not involved. The lesion extended downward in the same relative position about 1 cm. (2-5 in.) from this level. The upper part of the scar, in a section from a higher level, involved the posterior limb of the internal capsule, and as the island of

Reil is very short at this level the scar extended forward here beyond the anterior end of the island.

It will be noted that the lesions in this case involved both the external capsule, the anterior limb of the interior capsule, the lenticula, and to some extent the posterior limb of the internal capsule; also slightly the island of Reil. It is therefore difficult to draw any positive inferences with regard to the lenticula; the motor aphasia may have been due to lesion of the external capsule and the island, or to the lenticular lesion.

This man, several weeks after the beginning of his right hemiplegia, had slow speech; he apparently understood all that was said to him, but was not always able to find words to answer.

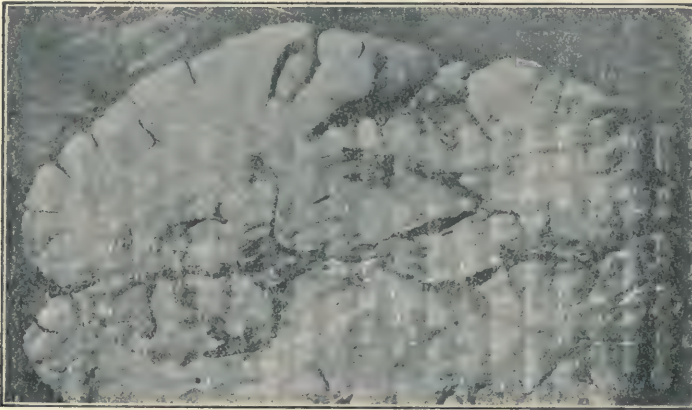


Fig. 8. A hemorrhagic cyst is present in the left lenticular nucleus. Some motor aphasia persisted. (Case 8.)

When the proper answer was repeated to him, he recognized it. There was apparently some motor aphasia, but it was by no means complete, and yet a large part of the putamen was destroyed.

The occurrence of the hemorrhage into the external capsule of one side is interesting in connection with what has been said of lesions thus located and of the anatomical and functional separation of the basal ganglia from the insula.

Case 9. Left Facial Paresis—Deviation of Tongue to the Right—Spasticity of Right Upper Limb and of the Lower Limbs—Babinski Response on Both Sides—Spasticity Present and Deep Reflexes Exaggerated on the Left—Incontinence of Urine and Feces—Involuntary Laughing and Weeping—Mentality Dull—Speech Indistinct but no true Motor Aphasia Present—A Small Cyst in the Left Putamen extending into the White Matter of the Insula.

R. S., sixty-two years old, white, German, was admitted to

the nervous wards of the Philadelphia General Hospital October 24, 1905.

Examination showed the left naso-labial fold to be less marked than the right. The tongue protruded to the right. The brow was wrinkled equally well on the two sides. The pupils were unequal, the left being larger than the right. The ocular movements seemed to be preserved, but the patient's mental condition rendered the examination of these movements a somewhat uncertain matter. On attempting to close the eyes, the left was not so well closed as the right, the edges of the left being separated by quite a little distance as compared with the right. The pupils reacted to light and in accommodation.

The patient seemed to have considerable strength in both upper extremities, all movements being preserved. A suggestion of spasticity was present on the right side. The reflexes of the right upper limb were increased; the left were not increased. The patient had lost control over the bladder and bowels. Movements of the legs were restricted and appeared to be spastic, but it was here again, as with the ocular movements, difficult to say how much was paralysis and how much failure to understand. Both knee-jerks were plus and the Babinski response was obtained on each side. The ankle jerk was also much increased on each side. The biceps jerks were present on both sides.

Under date of December 18th additional notes were made as follows: The right knee-jerk was plus; the left plus, plus; the ankle jerk was plus on the right side and also on the left; Gowers' front tap was present on the left side and absent on the right; ankle clonus was absent on both the right and the left sides.

Stimulating the sole of the foot gave very uncertain results. On the right side at times extension of the toes appeared to be present, and at other times, flexion. On the left side the tendency seemed to be towards the extension or Babinski response.

Power in both upper limbs was much diminished. Movements were much better performed with the right upper extremity than with the left. Fine movements were very poorly and clumsily performed. Movements were very ataxic, especially those of the right side. Marked tremor was present when the fingers of each hand were extended. The limbs on the right side were spastic, and contractures were present. The patient could move the right leg, but not nearly so well as the left. Sensation to touch, pain and temperature seemed to be undisturbed.

On December 27th further notes were made. The patient's condition was not so good as it had been. The nurse stated that the patient had occasional spells of laughing and crying; her mentality was not clear; speech was not distinct, and it was only with difficulty that she could be understood. She had no trouble with chewing or swallowing; she had no dribbling of

saliva. The pupils were unequal, both being larger than normal, the left larger than the right. Extraocular movements were normal. It was impossible to tell the condition of the light reflex and accommodation and convergence. Apparently the seventh nerve on each side was not involved. Apparently ptosis of the right upper lid was present. The other conditions were as noted in the preceding observations.

The bedside notes contain a statement without date made by Dr. Spiller that the patient's speech was very indistinct, but that she was not aphasic.



Fig. 9. A small cyst is present in the left external capsule, extending into the lenticular nucleus. The speech was indistinct, but not aphasic. (Case 9.)

On February 4, 1906, while eating her dinner, she suddenly turned to the attendant who was feeding her and said that she had enough. Immediately after this she gasped for breath, became cyanosed and fell over dead.

At the necropsy the pathological findings showed the presence of hypertrophy of the heart; chronic passive congestion of the spleen; chronic interstitial nephritis; passive congestion and fatty infiltration of the liver and general arteriosclerosis. The skull appeared to be thicker than normal. The brain and spinal cord were retained for subsequent investigation.

The gross lesion found was a small cyst 0.5 x 0.5 cm. (1-5x1-5 in.) in the middle of the left putamen in a horizontal section through the upper part of the basal ganglia. This cyst

did not extend into the cortex of the island of Reil, but implicated the white matter in the middle of the island.

It would seem in this case that the anarthric or dysarthric affection of speech and the impairment of power and spasticity and exaggerated reflexes on the right side might be fairly attributable to the lenticular and subcortical lesion. The speech disorder in particular is similar to that which seems to have been observed in other cases of limited lesion of the putamen—not a true motor aphasia, but a speech disturbance more nearly allied to that which is seen in pseudo-bulbar paralysis from capsular lesions. The woman was sixty-two, and some of the symptoms may have been caused by senile changes in the brain, but this finding is important, viz., a small cyst interrupting the fibers in the left external and extreme capsules, and extending into the putamen in about its middle portion.

Case 10. *Apoplectiform Attack without Unconsciousness—Speech Lost for Two Hours, then Recovered—Hemiparesis of Right Lower Face—Deviation of Tongue to Right—Paralysis of Right Upper and Lower Extremities—Reflexes increased on right—Sensation not impaired—Destructive Lesion involving the Left Lenticula and both the Anterior and Posterior Limbs of the Internal Capsule—Absence of Persistent Motor Aphasia.*

A. W., white, thirty-six years old, was admitted to the nervous wards of the Philadelphia General Hospital December 27, 1898. The family and previous history of the patient contained no fact of importance, except that her mother died of dropsy at the age of forty-two. The patient used alcohol moderately.

Five weeks before admission the patient had an attack of influenza from which she recovered. Three weeks before admission she was taken with weakness while on the street and felt three times. While unable in these spells to stand or walk, she remained perfectly conscious. Her speech was affected for two hours, during which time she was unable to talk. On attempting to move the right arm or leg she was unable to do so.

Examination showed a fairly well nourished adult. The pupils were equal; the irides reacted to light, and in accommodation and convergence. The extraocular muscles and the eyelids were not paralyzed. The muscles of the right side of the face appeared to be partially paralyzed, this being especially noticeable in the lower part of the face. The tongue deviated to the right on protrusion.

The right arm and leg were completely paralyzed, but there was no limitation of movement in the left extremities. The biceps and triceps jerks were increased on the right side and absent on the left; both knee-jerks were increased. Slight patellar clonus was present on the right side; ankle clonus was absent. The

plantar reflexes were increased on the right side. Sensation was nowhere impaired.

The pulse was frequent, small, and fairly strong. The heart sounds were normal; the second sound was accentuated.

Examination of the lungs showed nothing on palpation and percussion, but on auscultation at the right apex was heard a slight prolongation of expiration, especially noticeable at the middle lobe posteriorly.

An occasional note regarding this patient was made between her admission and November 17, 1903, simply showing that the patient was a right hemiplegic. On November 17, 1903, examination showed that the woman was weak and her breathing shallow, but that she was conscious. Both the right upper and lower extremities were completely paralyzed. Knee-jerks were present on both sides, prompt on the right; the Babinski response could be elicited on the right, but was absent on the left; neither ankle clonus nor patellar clonus was present. The tongue protruded slightly to the right. No old contractures were present on either side. Sensation was not impaired.

Notes were made at this date that the patient had not slept well. She was constantly muttering and picking at the bed-clothes. She could, however, be aroused and made to answer questions intelligently. Respiration was increased, but not labored. The tongue was heavily coated. Examination of the urine was negative. Resonance was impaired over portions of the middle and lower lobes of the lungs on the right side; breathing was distinctly tubular over an area of the middle lobe. On November 18th the patient developed edema of the lungs; the heart became very rapid and weak. She died November 19, 1903.

The following conditions were found post mortem: Croupous pneumonia with red hepatization and right-sided slight hydrothorax; slight chronic diffuse nephritis and acute parenchymatous nephritis; also chronic passive congestion of liver; slight fatty degeneration of liver; slight chronic thickening of the pia arachnoid; atheroma of thoracic and abdominal aorta.

A horizontal section was made through the left cerebral hemisphere, just at the upper border of the head of the caudatum and thalamus, so as not to remove any of these structures. At this level a cyst was found occupying the entire area of the lenticle and encroaching upon the anterior and posterior limbs of the internal capsule and the white matter of the island of Reil. The cortex of the latter region was intact. The head of the caudatum and the thalamus, so far as could be determined by the naked eye, were not implicated. The cyst did not extend forward or posteriorly beyond the limits of the lenticle. It measured 3.5 cm. in length and 1.5 cm. in width (1 2-5x3-5 in.).

A horizontal section made through the cerebral hemisphere

1 cm. (2.5 in.) below the previous section showed the lower termination of this cyst. It was directly beneath the center of the cyst as shown in the previous section and measured 1.5 cm. long by 1 cm. wide (3.5x2.5 in.). Sections of the spinal cord made by Dr. Weisenburg showed intense degeneration of the right crossed and left direct pyramidal tracts.

The lesion present in this case, although in large part in the lenticula, was not confined to it, extending both into the anterior

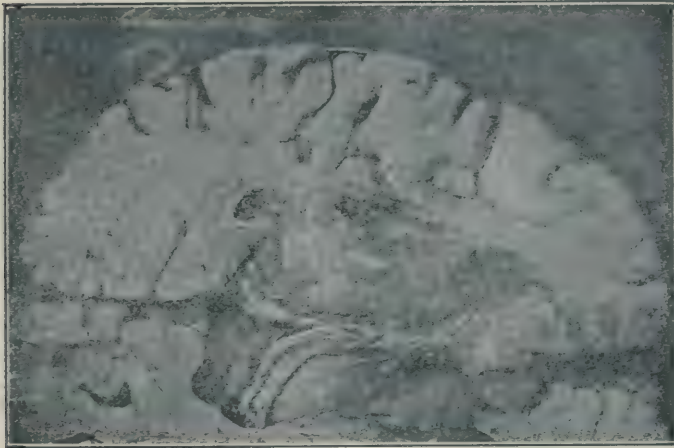


Fig. 10. Complete destruction of the left lenticular nucleus and anterior and posterior limbs of the left internal capsule, without persistent motor aphasia. (Case 10.)

and posterior limbs of the internal capsule. We cannot therefore draw any positive conclusions as to the part played by the lenticular lesion in the production of the hemiplegia. The case is chiefly of value in throwing some light upon the effect of so large a lenticular lesion on the function of speech. Speech, while affected at first, was soon recovered, and the patient continued to speak intelligently. She was not a motor aphasic.

This in connection with the first case of this paper shows that complete destruction of the left lenticula does not necessarily cause complete and persistent motor aphasia. While the notes on the condition of speech in this case are not as complete as might be desired, they indicate very clearly a recovery, in part at least, of the motor speech function, and we have the statement from Dr. T. H. Weisenburg that when a resident in the hospital he frequently conversed with the patient, and that she was able to speak. We cannot conclude, however, that speech was normal.

Case 11. *Aploplectic Attack causing Complete Right Hemi-*

plegia with Hemianesthesia and Complete Aphasia—Disappearance in large part of Speech Affection and Hemiplegia—Persistence of Hemianesthesia—Right Lateral Anopsia—Dyslexia—Partial Word Deafness and Partial Word Blindness—Deviation of Tongue to the Right—Right-sided Spasticity and Contractures and Exaggerated Reflexes—Large Cyst involving Left Extreme Capsule, Claustrum and External Capsule and Destroying the Left Lenticula and Carrefour Sensitif, also involving Optic Radiations—The Anterior Half of the Posterior Limb of the External Capsule and the Anterior Limb of the Internal Capsule not involved.

A case which was first recorded by one of us, Dr. Mills,²⁸ in a paper which was written in collaboration with Dr. G. E. de Schweinitz, is of some value in connection with the study of the motor and speech functions and the question as to the sensory functions of the lenticula. This patient died in the wards of Dr. F. X. Dercum, in the Philadelphia General Hospital, and the case was made the subject of a paper by Drs. Dercum and Spiller,²⁹ on permanent hemianesthesia resulting from destruction of the *carrefour sensitif* and the lenticula without implication of the thalamus, the case appearing to demonstrate that the cerebral sensory fibers are located chiefly, if not altogether, in the area of the *carrefour sensitif*, or it may be that some sensory fibers pass through the lenticular nucleus, as held by Edinger. The writers define the *carrefour sensitif*, following Verger, as comprising the last third of the posterior limb of the internal capsule in the most anterior portion of the opto-striate region.

The hemorrhagic apoplectic attack which left the cyst found at necropsy occurred many years before the death of the patient. Immediately after the attack the man was completely paralyzed in the right arm and leg, and also completely aphasic and hemianesthetic. He recovered his speech very largely in about two months and the motor paralysis largely disappeared in about three months. The hemianesthesia remained and was extremely marked up to the time of the patient's death. The man was paralyzed in 1892. Four years later, in 1896, when the clinical report of Dr. Mills and Dr. de Schweinitz was made, he had right lateral quadrant anopsia; absence of Wernicke's symptom; dyslexia; right hemiparesis; partial right hemianesthesia; and partial word deafness and word blindness. His tongue was slightly tremulous on protrusion and deviated a little to the right. With regard to his aphasia, the patient stated at

²⁸Mills, Chas. K., and de Schweinitz, Geo. E. The Philadelphia Hospital Reports, Vol. III., 1896.

²⁹Dercum, F. X., and Spiller, William G. The Amer. Journ. of the Med. Sciences, N. S., 123, March, 1902.

this time (1896) that from the first he knew what he wanted to say, but could not put it into such words as would be understood by his hearers. He could recognize objects, but could not pronounce their names. He could not read writing or print, because everything ran together; he recognized the letters, but could not pronounce them. He understood sentences, but could not read them. In 1896 he had almost completely recovered from his aphasia, although his voice was a little thick and difficult to understand.

In 1899 there was present a spastic hemiplegia of the right



Fig. 11. A. Portion of the lenticular nucleus not destroyed. B. Anterior limb of the internal capsule. C. Head of the caudate nucleus. D. Posterior limb of the internal capsule. E. Carrefour sensitif. F. Cyst in the lenticular nucleus. (Case 11.)

side, moderate in severity, with a moderate degree of contracture of the right arm and spastic rigidity and exaggeration of the knee-jerk on the right side. The right side of the face was only slightly involved. The hemianesthesia appeared to be everywhere complete, and was associated with right-sided homonymous hemianopsia.

"A cavity, probably resulting from an old hemorrhage, 2.8 cm. in depth from above downward, and 2.5 cm. in its longest diameter, (1 1-8 in. x 1 in.), was found in the extreme

capsule, claustrum and external capsule of the left side. The left lenticula appeared to be almost destroyed and the extreme posterior part of the posterior limb of the internal capsule and the optic radiations were involved in the destructive lesion. The anterior half of the posterior limb of the internal capsule was not implicated in the cyst. The anterior limb of the internal capsule was entirely normal. The thalamus was not implicated at all in the cyst, and the external medullary lamina of the thalamus was distinct and normal in appearance. The external nucleus seemed to be of about normal size. A transverse cut passing through the hypothalamic region showed the anterior half of the posterior limb of the internal capsule apparently normal. The lenticular nucleus, except in its extreme ventral portion, was destroyed.

"The portion of the thalamus that showed atrophic change was the pulvinar, and this alteration was the result of the destruction of the optic radiations."

In addition to the macroscopical examination of the specimen in this case, careful microscopical investigation by serial sections was also made, this confirming the fact that the anterior half of the posterior limb of the external capsule and the entire thalamus were not involved in the lesion, while the *carrefour sensitif* and the lenticula, with the exception of its most ventral portion, were destroyed.

It would seem fair in this case to refer the moderate spastic hemiparesis which persisted until the death of the patient partly to the lenticular lesion, as the motor subdivision of the posterior limb of the internal capsule was only a little involved. The early aphasia which disappeared in two months was probably due to compression or partial destruction of some part of the lenticula concerned in speech phenomena, which must be the anterior portion, the compression of the neighboring speech areas in the temporal lobe and to the destruction of the insula. We are inclined to refer the permanent hemianesthesia chiefly to destruction of the *carrefour sensitif*, although we do not deny that sensory fibers may pass through the lenticula. The disturbance of both phonation and articulation which is referred to in the statement that his voice was thick and difficult to understand, was probably dependent upon the lenticular lesion, as the genu and anterior half of the posterior limb of the internal capsule were not destroyed.

Both from our study of the literature of the subject and from the analysis of the personal cases given in this paper, we find it is somewhat difficult to form positive conclusions as to the symptomatology of lesions of the lenticular zone.

We believe, however, that a few general conclusions may be drawn:

1. Lesions restricted to the lenticula apparently do not cause sensory symptoms;
2. Motor symptoms probably result from lesions situated in certain parts of the lenticula; speaking generally, the lenticula may be regarded as a motor organ;
3. Anarthric or dysarthric speech disorders result from lesions of some portion of the left lenticula, which probably contains centers which are concerned with the movements which make speech possible;
4. Destructive lesions of certain portions of the lenticula probably cause a paresis of the limbs or face;
5. The paresis or paralysis caused by destructive lesions of the lenticula differs from that produced by capsular lesions, the impairment of power not being so severe and not being so characteristic in the former as in the latter case;
6. The paresis or paralysis which is caused by lenticular lesions differs from that produced by cortical lesions in that it is less likely to be dissociated; although dissociated lenticular paresis may occur;
7. While the loss of power which results from a destructive lenticular lesion is permanent, it is usually not intense;
8. Persistent true motor aphasia, as this form of speech disorder is generally understood, is not caused by a lesion restricted to the lenticula, no matter what its size or destructiveness;
9. The insula, cortex and subcortex play an important part in speech phenomena, one entirely different from that played by the lenticula and the internal capsule;
10. The insula is a part of the cortical motor center for speech, Broca's convolution probably forming with the insula the entire cortical motor center for speech;
11. Motor aphasia may be present without a lesion of the left third frontal convolution;
12. The lenticula forms too large a portion of the cerebral hemisphere to be regarded merely as a vestigial organ.

GLIOMATOSIS OF THE PIA AND METASTASIS OF GLIOMA.*

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Various forms of malignant tumor may implicate the pia of the spinal cord and brain diffusely. The sarcomatous infiltration is the best recognized. The first case of this kind reported in America, so far as I know, is that of George K. Weaver,¹ in 1898. In 1903 I² reported two cases, one in association with W. F. Hendrickson, and the present communication is made in relation to one of these. Since then two cases have been recorded by F. X. Dercum.³ The German literature contains the largest number of cases and a few may also be found in the British journals.

Sarcomatous infiltration, however, is not the only form of diffuse tumor formation of the pia. The carcinoma and glioma have been known to appear in this manner. An important paper in regard to gliomatosis of the pia has recently been published by Grund.⁴ This author says that the earliest mention of glia proliferation in the pia is by Klebs, who found glia in the pia near a glioma of the brain. He cites Alzheimer as authority for the statement that glia proliferation in the pia is common in parietic dementia. Schlesinger and Saxer have seen proliferation of glia in the pia in syringomyelia. Schlesinger, in his monograph on tumors of the spinal cord, makes no mention of diffuse glioma in the pia arising from the neuroglia, but since the publication of his monograph four cases have been reported (Leusden, Fränkel and Benda, Fischer, Roux and Paviot) and Grund adds one more, but does

¹Weaver. *Journal of Experimental Medicine*, 1898, Vol. III., No. 6.

²Spiller and Hendrickson. *American Journal of the Medical Sciences*, July, 1903.

³Dercum. *Philadelphia Hospital Reports*, Vol. V., 1903, and *JOURNAL OF NERVOUS AND MENTAL DISEASE*, March, 1906, p. 169.

⁴Grund. *Deutsche Zeitschrift für Nervenheilkunde*, Vol. XXXI., Nos. 3 and 4, p. 283.

*Read before the Pathological Society of Philadelphia, Feb. 14, 1907.

not refer to my case, so that at the present time six cases of gliomatosis of the pia are on record.

The glioma, unlike the sarcoma, carcinoma, endothelioma or perithelioma, is not a sharply defined tumor of the nervous system, but infiltrates into the surrounding tissue, and it is owing to this fact that its surgical removal is so difficult. Indeed, I⁵ have advocated leaving the tumor untouched if at the operation it can be shown to be a glioma. In the striking language of Walton and Paul,⁶ one might as well attempt to remove the decayed spot from a fruit in order to arrest the process of destruction as attempt to remove a glioma in order to arrest the tumor formation. It may be possible, however, when the glioma is very minute, to remove surgically sufficiently surrounding brain tissue to prevent further progress of the growth.

Unlike sarcoma, glioma seldom extends into the membranes of the brain or cord, and never breaks through beyond the membranes. Glioma, unless very cellular and closely resembling sarcoma, grows very slowly. Max Borst,⁷ in his work on tumors, has a very interesting chapter on glioma, and according to him cavities in a glioma lined with cylindrical cells are rare, but in this finding the embryonal relation of the glia cells to the ependyma may be recognized. Glia cells as derivatives of ependymal cells may have the power to return to the type of epithelium of the primary neural canal, or the cylindrical cells in the glioma may be regarded as separations from the ependymal cells of the ventricles from which the glia cells of the tumor are derived. Both conditions are possible.

The term gliosarcoma has fallen into some disrepute, as it implies an origin from both mesoblastic and epiblastic tissues, but Borst employs the term for the mixed tumor and calls the glioma rich in cells glioma sarcomatodes. The distinctions between glioma and sarcoma may be difficult to make. The presence of glia fibers does not make a tumor a glioma, especially if the fibers are near the border of the tumor, as

⁵Spiller and Frazier. University of Penna. Med. Bul., September, 1906.

⁶Walton and Paul. JOURNAL OF NERVOUS AND MENTAL DISEASE, August, 1905, p. 481.

⁷Borst. Die Lehre von den Geschwülsten, Vol. I.

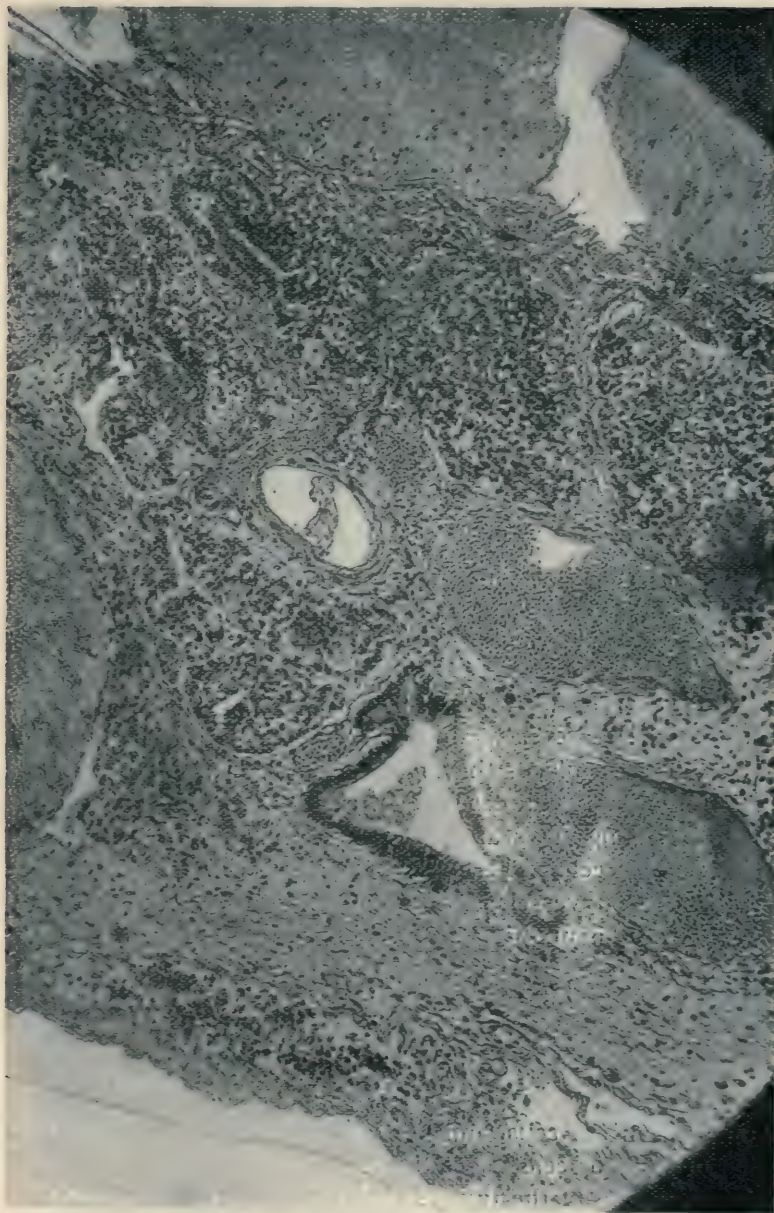


Fig. 1. Diffuse tumor formation of the pia of the spinal cord with rows of ependymal cells inclosing spaces.

they may be the remains of neuroglia caught within the tumor, and glia fibers, Borst states, occur in almost every growing tumor of the central nervous system. Sometimes what are supposed to be glia fibers are really fibrin fibers, and these may closely resemble the glia fibers, and they have no connection with the glia cells. Borst even goes so far as to say that it is uncertain whether a glioma may not be composed entirely of cells, *i. e.*, may be without fibers, and he implies that such a form of glioma may occur, the differentiation of such a tumor from a sarcoma might therefore be impossible.

In a case reported by me in 1903, to which reference already has been made, very many minute tumors and tumor infiltration were found in the pia of the spinal cord. The structure of this tumor infiltration of the pia was so extraordinary that I hardly knew what name to give to it. It resembled closely tumor arising from the ependyma, (See fig. 1.) but I could find no record at that time of gliomatous infiltration of the pia, and none of the eminent pathologists I consulted had ever heard of such an occurrence. I recorded the findings as follows:

"The tumors everywhere have much the same structure. They consist of round or somewhat elongated cells, and even by Mallory's neuroglia stain contain very little intercellular tissue. In some places the cells are distinctly columnar and resemble those of the ependyma. The columnar cells are arranged in long rows and have a large nucleus situated at one end of the cell. These rows of cells are found especially about the blood vessels, but also where there are no blood vessels. There is unquestionably a close resemblance between these cells and those of the ependyma, and the temptation, therefore, is to call the tumor an ependymoma, but it seems remarkable that an ependymoma, being a form of glioma, should give extensive metastasis to the pia of the spinal cord, which is of mesodermic origin. This, and the absence of glia fibers between the cells, and the distinct tendency to the formation of rows of cells about the blood vessels, seem to justify the classification of the tumors under the sarcomata, possibly endotheliomata or peritheliomata; and yet the close resemblance of some of these cells to those of the ependyma may permit us to regard the growth as a mixed one, and as being

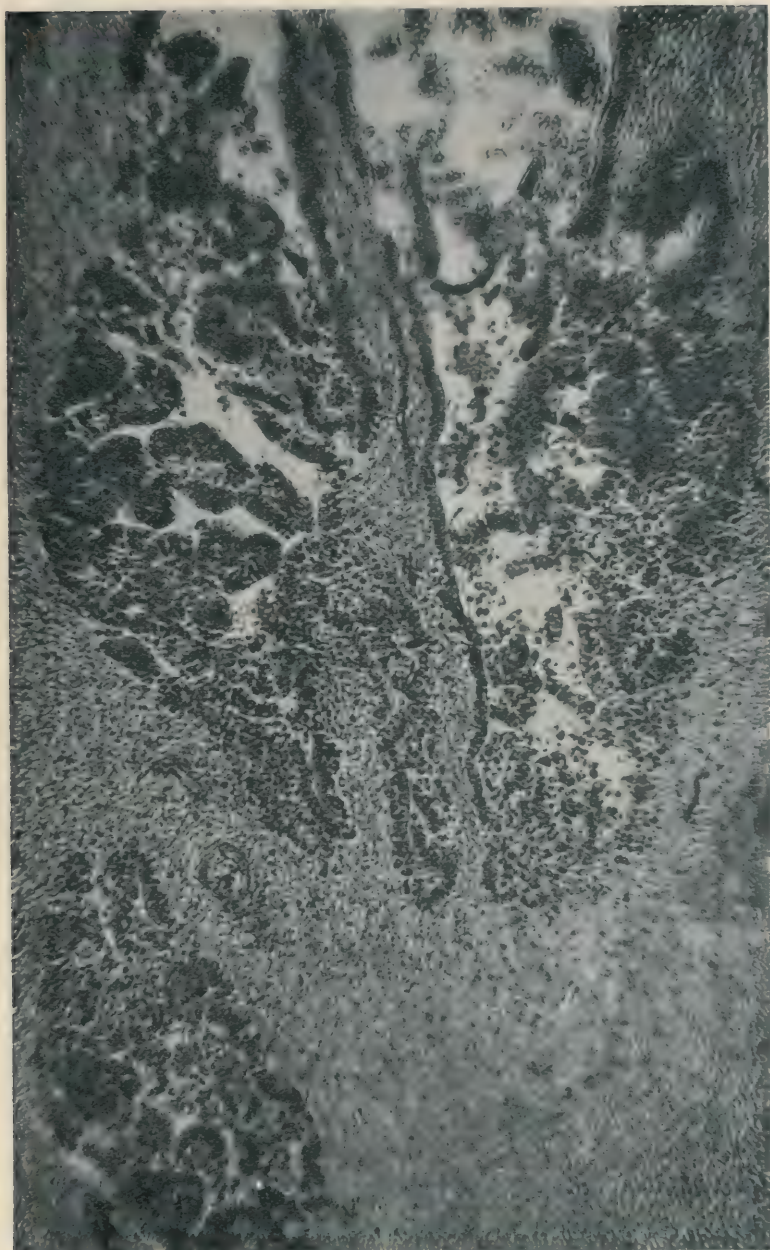


Fig. 2. Tumor filling the fourth ventricle. The proliferation of the ependymal lining is clearly shown.

partly a sarcoma and partly an ependymoma. The ependyma is known to proliferate in cases of syringomyelia when the cavity extends to the central canal and to cover in part the wall of the cavity. It is not unreasonable to suppose that a sarcoma growing from the pia may extend to the fourth ventricle, and by irritation of the ependyma lead to its proliferation, and cause in this way a mixed tumor." I referred to the fact that the tumors of the pia in this case had a resemblance to the neoplasms described by Rosenthal, Fränkel and Benda.

Since the publication of this paper the possibility of diffuse gliomatosis has been in my mind, and I have been waiting for further evidence of its occurrence, and this is now given in Grund's paper. I am now inclined to regard the sarcomatous nature of the tumor infiltration in my case as doubtful, and I look upon the growth as multiple gliomatosis. A section from the medulla oblongata shows a tumor filling the posterior part of the fourth ventricle, and the proliferation of the ependymal cells lining this ventricle is so pronounced that the ependymal origin of the diffuse process seems fully demonstrated. (See fig. 2.) These cylindrical cells no longer form a row lining the ventricle, but occur in long rows, or bordering papillomatous processes, clearly arising from the ependyma, and extending outward into the tumor.

In another case, reported by me⁸ briefly from a clinical aspect and with scarcely any reference to the pathological findings, a tumor filled the posterior part of the fourth ventricle and extended downward upon the cord as low as the sixth cervical segment. This tumor also contained rows of cells resembling those of the ependyma, and forming the lining of large irregular spaces. It was an ependymoma. Another, but smaller tumor, having the same structure, was found upon the lower part of the thoracic region of the cord and was probably a metastatic growth, and therefore exceedingly interesting. Borst says that metastasis of glioma is exceedingly rare, and that Stroebe in one case saw regionary metastasis, *i. e.*, near the original tumor. Multiple gliomata, according to Borst, have not positively been observed.

⁸Spiller, Musser and Martin. University of Penna. Med. Bul., March and April, 1903.

PRESERVATION OF DEEP SENSIBILITY OF
THE FACE AFTER DESTRUCTION OF
THE FIFTH NERVE.¹

BY R. H. IVY AND L. W. JOHNSON,
Class of 1907.

RECENT observations and experiments in regard to the functions of the nerves have tended to lead us far away from the conceptions which were held a decade ago concerning the forms of sensation of individual nerves. These studies have been made possible in large part by the great advances in our knowledge and in surgical technique in operations on the nerves and their ganglia. Thus, most of the cases reported as studied have been cases in which nerves were cut as a therapeutic measure, or for purposes of experiment; a smaller number have been those in which a functional loss of the nerve has been brought about by a neoplastic growth or similar factor.

The great amount of literature bearing on this broad subject, and the very numerous phases of it which have been studied, make it necessary that a paper so modest in its scope as the present one should present only a few considerations of the many which are so pertinent to the question.

This paper, therefore, is concerned chiefly with the conduction of sensory impulses from the face to the sensorium, and especially the conduction of deep pressure sensibility

¹ Read before the Philadelphia Neurological Society, April 23, 1907

as contrasted with that of cutaneous sensibility to light touch, temperature, and pain sense. Up to a comparatively recent period it was considered that all sensory impulses of the face were carried by way of the fifth nerve, while the seventh nerve was concerned solely in the motor supply of the muscles to which it runs. Recently, however, the opinion has been gaining ground that we must modify this teaching to a considerable extent; and we purpose, in this paper, to present some facts in support of the changed conception of the nerve supply of the face.

Very few observers have an opportunity to study cases of uncomplicated lesions of the fifth nerve, with subsequent verification by necropsy; and, furthermore, it is exceptional that intracranial growths cause so little involvement of mentality as to permit of the delicate sensory tests required in this kind of investigation.

Head,¹ in 1905, published the results of his observations on sensory disturbances produced by section of sensory nerves of the forearm and hand. The portions of his findings which may bear on the present subject are as follows:

Complete division of the median and ulnar nerves at the wrist abolished sensations to prick, light touch, and changes in temperature, but not to pressure. "Cotton-wool rolled up into a pledget applied to the skin with some force produced a definite sensation; but if the skin was lifted the same method of stimulation entirely failed to evoke any response, a proof that whatever sensation had been previously present was due to the integrity of the underlying structures. Thus, if a part is deprived of all its cutaneous nerves it becomes entirely insensitive to prick, to touch, to all forms of temperature; but it still remains sensitive to any stimulus which jars the skin, however lightly." If, in addition to the nerves, the flexor tendons at the wrist are divided, pressure can no longer be appreciated over the area insensitive to prick, showing that pressure sensation is conveyed by fibers running in the tendons.

Head also mentions Sherrington's demonstration of afferent fibers running in the nerves which supply the muscles, and of the fact that section of motor nerves deprives the muscles and deep structures of all sensation.

From these facts Head concludes that "deep sensibility, capable of answering to pressure and to the movement of parts, and even capable of producing pain under the influence of sufficient pressure, is conveyed through afferent fibers running in the motor nerves."

The demonstration of these peculiarities of the sensory nerve supply of the trunk and limbs suggests the application of the same principles to the sensory nerve supply of the face. The idea that the seventh nerve in some way or other contains sensory fibers is not new, and tumors of the Gasserian ganglion or operations on the fifth nerve have made it possible, by eliminating the fifth, to study the relation of the seventh nerve to certain forms of sensation.

Spiller,² on April 24, 1906, reported before the Philadelphia Neurological Society 2 cases in which the Gasserian ganglion had been removed, the deep pressure sense in the area of of the fifth nerve remaining, while all other forms of sensation were lost. (Dana's article appeared in September of the same year.)

The importance of these investigations is clearly shown as regards the surgery of the fifth nerve. It has been noticed, not infrequently, that after operation on the Gasserian ganglion or the sensory root of the fifth nerve some sensation has been retained in the side of the face supplied by the nerve operated upon. When this has occurred, it has at times been assumed that the operation was imperfect. Two fallacies are to be considered in this connection. It is frequently forgotten that the fifth nerve supply does not extend all the way to the border of the lower jaw, but that a considerable area over the lower part of the cheek receives its sensation by way of the cervical nerves. The second fallacy is in regard to pressure sense; as when touch is associated with pressure,

even when the latter is but slight, the tactile sensation may seem to be present when in reality it is the pressure sense that is retained. The assumption that the Gasserian ganglion has not been destroyed or the sensory root cut is more readily accepted in such cases of partial preservation of sensation on account of the well-recognized difficulty of operation on the intracranial portion of the fifth nerve. The subject is important also because after surgical intervention on the fifth nerve a certain form of sensation may be preserved so that there is not complete anesthesia even with complete destruction of the fifth nerve. In making tests we must also consider the normal variations in acuteness of perception of sensibility of the two sides of the body which occurs to a greater or lesser degree in all persons.³

Many writers⁴—Van Gehuchten⁵ says most writers—now describe the facial nerve as in part sensory and in part motor. Retzius, and more recently von Lenhossèk,⁴ have shown that the geniculate ganglion is like the cerebrospinal ganglia, and that the central processes of its cells pass into the nerve of Wrisberg. Van Gehuchten finds that at its exit from the stylo-mastoid foramen the seventh nerve contains undoubtedly a certain number of sensory fibers which arise in the geniculate ganglion. The nerve of Wrisberg is therefore the sensory root of the seventh nerve.

Amabolino⁵ (cited by Van Gehuchten) has formed a different opinion. He believes that the peripheral processes of the cells of the geniculate ganglion pass into the chorda tympani and that none of them pass into the peripheral branches of the seventh. Van Gehuchten points out that Amabolino found about one-fifth of the cells of the geniculate ganglion normal after section of the chorda tympani, and very pertinently asks where these cells send their processes if not to the peripheral branches of the facial. Biehl⁶ argues that the retention of sensation after operations on the fifth nerve is not due to regeneration of this nerve, or to an ingrowing of fibers from the fifth nerve on the opposite side

of the face, or from cervical nerves, as it appears too soon after the operation, also, the limits of the area in which sensation is preserved after removal of the Gasserian ganglion correspond quite well with the limits of the area of disturbed sensation in the case of facial paralysis. Dana,⁷ after testing a series of cases with lesions of the fifth nerve, comes to the conclusion that, "however true the views may be as to differentiation of afferent nerves into two kinds as applied to the body and limbs, it does not seem to apply to the head. It would seem that it is confined to the specific arrangement of spinal nerves, while the central nervous system and cranial nerves conduct sensations in a different way." To support this view he details a case in which the third division of the fifth nerve was cut as it issued from the dura, without involving any other branch. The result of this was an area of anesthesia involving the upper lip and a portion of the inside of the nose and cheek, radiating upward toward the ear halfway and within about an inch of the eyelid; also anesthesia of the mucous membrane of the mouth and tongue, of the whole inner surface of the mouth on the left side, including the gums of the upper and lower jaws and the palate to the edge of the soft palate. The area was tested by cotton, by light stroke of the camel's hair brush, by pricking with a pin, by pinching, by application of ice and very hot water in tubes, also for sense of position and deep muscular sense. An absolute loss of all these forms of sensibility was found, no matter how hard the pressure. There was no anesthesia of the ear or meatus; there was some ataxia of the muscles of the face on the affected side; he could draw the mouth to the left, but had poor control. There was also anesthesia of the periosteum of the gums. He could not tell when the mouth was open.

In view of the distinct statement that the third division of the fifth nerve was cut cleanly, without severing any other branch, it is difficult to harmonize the area of anesthesia which followed the operation with our conception of the

cutaneous area supplied by the third division of the fifth nerve. The area of anesthesia, as described and pictured, encroaches deeply on the area generally ascribed to the infra-orbital and the branches supplying the nose from the second division of the fifth nerve, while the region ascribed to the auriculotemporal branch of the third division of the fifth showed no loss of sensation of any kind. The fact that there was no anesthesia of the ear or meatus seems to uphold the statement of Hunt⁸ that after removal of the Gasserian ganglion and cervical nerves there still remains an area in the auricle and the external auditory canal in which sensation is preserved because its sensory nerve supply is from the seventh through the geniculate ganglion. The statement that the patient could not tell when the mouth was open is also difficult to explain. Inasmuch as the lower jaw on the left is continuous with the lower jaw on the right, forming in fact one bone, he could not open his mouth on one side without opening it on the other side also. Granting that there is loss of sense of position of the muscles attached to the bone on the affected side, will not the muscles of the sound side be moved also and give a knowledge of the position of the lower jaw-bone, whether the mouth be open or shut?

The cervical nerve supply on the affected side would also aid in giving the sense of position of the jaw.

Dana also describes a case in which a very careful examination was made of the sensory condition after complete paralysis of the seventh nerve; it apparently showed that with paralysis of the seventh nerve there was no loss of deep sensation or sense of position.

The difficulty of studying the deep sensibility of an area where the cutaneous sensibility is not disturbed must lessen somewhat the value of this case as an argument that cutting the seventh nerve produces no loss of deep sensation. Head¹ states that deep sensation is best studied in a part where the skin is totally analgesic by reason of section of the sensory

nerves supplying the part. Dana has not fulfilled this postulate, but has assumed that there was no loss of pressure sense because the patient noticed no impairment on testing through the skin supplied with sensation through the fifth nerve. The patient must be a trained and careful observer who can detect the presence or absense of the seventh nerve impulses while the fifth nerve is still intact.

The case which has given us the impetus to study sensation of the face, briefly reported, is as follows:

A. K., male, aged twenty-five years; University of Pennsylvania Hospital, March, 1907, in the service of Dr. Spiller.

The patient had considerable stupor, but was able to respond intelligently to all questions. He was able to stand and walk, but had some ataxia of gait and station. Both eyes showed a high grade of choked disk. The left pupil was considerably larger than the right; neither pupil responded to light. Left eye: This showed slight divergence. The function of the external rectus was normal; movements of the internal, superior, and inferior recti were markedly limited. No wheel movement was obtained. Right eye: In this there was loss of movement of the external rectus, and marked limitation of movement of the superior and inferior recti, function of the internal rectus being almost lost. There was complete motor and sensory paralysis of the right fifth nerve supply, which will be described in full a little later. The cause of this paralysis was a tumor growing from the dura of the right middle cranial fossa, having its centre in the location of the right Gasserian ganglion, infiltrating the dura of a large part of the middle cranial fossa, completely destroying the Gasserian ganglion and involving the motor and sensory roots of the fifth nerve to their exit from the pons; infiltrating also the dura over the central part of the petrous portion of the temporal bone, in the right posterior cranial fossa, and extending over the basilar process of the occipital bone far enough to catch the left third nerve as it passed forward into the left middle fossa.

There was no question as regards the total destruction of the right fifth cranial nerve in this case.

Sensory disturbance in the area of the fifth nerve supply was as follows: Tactile sense was completely lost over the entire distribution of the fifth nerve on the right side, *provided no pressure was made*; there was loss of pain sense over the same area. Sensation was preserved along the border of the lower jaw in the area supplied by the cervical nerves. There was anesthesia of the cornea and conjunctiva on the right, with normal sensation of the left cornea and conjunctiva. A piece of paper put up the nose on the right side was not felt unless pressure was produced thereby, and did not cause lachrymation. The jaw went distinctly to the right when opened. Over the entire right side of the face, in the area completely insensitive to light touch, slight pressure was always recognized by the patient with his eyes closed, showing that the sense of pressure, even to slight stimulus, must have been produced through some other nerve supply. Owing to the condition of the patient, we were unfortunately unable to test pressure sensibility over the temporal region. The absence of pressure sensibility in this area would have been proof that this form of sensation is carried in the motor nerve to the underlying muscle, in this case the motor branch of the fifth. We also made no test of sensation in the cutaneous zone of the seventh nerve in the auricle and external auditory meatus, described by Hunt.⁸

In another case under Dr. Spiller's care a year ago the sensory root of the fifth nerve was cut because of the great pain from a tumor involving the Gasserian ganglion. The destruction of the fibers was made certain by the necropsy; and following this by microscopic study, intense degeneration of the sensory portion of the fifth was shown. The sense of touch was entirely lost over the area of distribution of the fifth nerve, while the sense of pressure was retained over the whole area. If there had been any doubt of the complete involvement of the fifth nerve by the tumor, the lesion was

made complete by the operation on the ganglion, which was followed by entire loss of cutaneous sensation over the area of all three branches of the fifth nerve.

So that in all Spiller has observed 3 cases in which pressure sensibility was retained after destruction of the Gasserian ganglion.

[NOTE.—The importance of the lacrymal reflex in diagnosing between organic and hysterical anesthesia of the trigeminal nerve has been pointed out by Spiller.⁹ Irritation of the nostril with a probe is the most satisfactory way of testing the reflex. While the lacrymal reflex in testing by this method may be lost, yet it is possible that it may be evoked by exposure of the eye to strong light.]

Cryer has reported 3 cases of peripheral operations upon the inferior dental nerve for trifacial neuralgia, in which pain on the inner surface of the lower jaw and floor of the mouth was not relieved by section below the point at which the mylohyoid branch was given off. A second operation above the point of junction of this branch with the inferior dental nerve was followed by complete relief. This result is evidence that the mylohyoid branch, which supplies motor fibers to the mylohyoid and digastric muscles, also conveys sensory impulses from these muscles, which from some irritation had given rise to pain.

Besides the conduction of deep sensation from the muscles to which it affords motor fibers, the facial nerve is also assigned a cutaneous zone which is described by Hunt⁸ as being situated within the auricle and external auditory canal. Extirpation of the Gasserian and cervical ganglia destroys the sensibility of the region about the ear, but there still remains this area in the interior of the auricle and the external auditory canal in which sensation is preserved.

Clinical, histological, and embryological investigations have shown conclusively that the facial is a mixed nerve (Hunt,

loc. cit.). Its afferent or sensory portion is the nerve of Wrisberg. Its ganglion is the geniculate, analogous to the spinal ganglia of the posterior roots in structure; its cell type corresponds exactly to that of the spinal and Gasserian ganglia.

From the foregoing cases and facts the evidence is strongly in favor of the sensory distribution of the face being conveyed in the same way as in other parts of the body.

Our conclusions may be summed up as follows:

The fifth nerve conveys fibers of sensibility to light touch, changes in temperature, and pain from those parts generally recognized as being within the area of distribution of the said nerve.

The seventh nerve conveys fibers of deep or pressure sensibility from the muscles which it supplies with motor fibers.

The muscles of mastication, which derive their motor supply from the fifth nerve, probably are also furnished with fibers of deep sensation by that nerve.

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THE OCCASIONAL LONG DURATION OF BRAIN TUMOR.

WITH THE REPORT OF A CASE OF JACKSONIAN EPILEPSY OF EIGHT
YEARS' DURATION AS THE ONLY SIGN OF A SMALL
CEREBRAL GLIOMA.¹

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REMARKS BY DR. SPILLER.

THE existence of symptoms of cerebral tumor during a period of eight years, with the finding at the end of that time of a tumor 1.5 cm. by 1.5 cm. by 0.5 cm., is so remarkable that the following case seems worthy of report:

Woods, aged sixty years, entered my service in the Philadelphia General Hospital June 17, 1904.

On examination after admission he wrinkled his forehead well on each side. The pupils were equal and regular in outline and reacted to light and in accommodation and convergence. The movements of the eyeballs were good. The tongue was protruded in the median line.

He was seen June 18 by Dr. Weisenburg, when the following notes were made: He had been well until eight years previously, at which time on one occasion he felt dizzy, and suddenly had twitching in his left fingers, then the

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twitching extended to the left shoulder and down the left side. He does not know how much time elapsed before he had a second attack. He had been getting worse during the past four or five years. His mentality is affected and he has some failure of memory. He has had many convulsions during the past four or five years, and the night before admission had convulsions all night. He knows when they are coming on because he is "worked up," as he expresses it, before they appear. They are confined to the left side. The nurse states that he has had many convulsions since he has been in the hospital, and that they have all been confined to the left side. The patient says that after the convulsions he loses power in the left limbs, especially in the lower limb, and that the limbs feel "numb and dead." The sensation of the left limbs is never normal, and since his first convulsion eight years ago the left limbs have not been so strong as they were previously. He has headache and dizziness when he has convulsions, but never vomits. His sight, he thinks, is good. He denies all venereal infection and says he drank little.

No impairment of stereognostic perception is detected, and sense of position is normal, as are also sensations of touch and pain in both upper limbs. The left limbs are not wasted. The masseter muscles contract normally.

He was seen by me June 20, when the following notes were made:

In a spasm observed by me both upper limbs became rigid, the face was drawn forcibly to the left, and the eyeballs also were drawn to the extreme left. The eyelids were closed. In about a minute the tonic spasm of the left side of the face and left upper limb gave place to clonic spasms; the left lower limb also was thrown into clonic spasms. So soon as the clonic spasms began on the left side of the body the right upper and lower limbs became partially relaxed. No clonic spasms were seen on the right side of the body or face. The tongue was not drawn to either side. The attack lasted one or two minutes.

In another attack the spasms were similar to those already described. The eyelids on each side were thrown into clonic spasms and the convulsive movements were equally intense in the lids of each side and in each side of the forehead. There were no clonic spasms of the muscles of the eyeballs. The right side of the lower part of the face was not implicated in the clonic movements.

The wrinkles of the forehead may not be quite so deep on the left side as on the right when the man looks upward. The left upper eyelid is more easily raised by the examiner than the right. When he attempts to smile the left nasolabial fold is not quite so deep as the right. He does not draw up the left corner of the mouth so well as the right. Sensation of pain is preserved in each side of the face, but the mental state does not permit an examination of tactile sensation. The tongue is protruded straight, is not atrophied and shows no fibrillary tremors. The man obeys simple commands and seems to understand what is said to him, although he is not very intelligent.

The left upper limb is very weak and he is unable to move any part of this limb, but the weakness was not so great yesterday. The biceps tendon reflex is almost abolished on the left side. The left triceps reflex is present, but is not very prompt. Pin-prick seems to be felt promptly in the left upper limb. It is impossible to get the patient to fix his attention on the examination of tactile sensation. The movements of the right upper limb are free at all parts and the grasp of the right hand is good. The biceps tendon and triceps tendon reflexes are not very distinct on the right side. He cannot recognize today any objects placed in the left hand, although he recognizes them promptly when they are placed in the right hand.

The movements in the right lower limb are good, but the left lower limb is completely paralyzed. The patellar reflex is present on each side, and distinctly prompter on the left side. Ankle clonus and Achilles tendon reflex are not present on either side. The Babinski reflex is distinct on the left side, the big toe being moved upward, but is not so distinct on the right side. Pin-prick is felt distinctly in each lower limb.

In a third convulsion seen by me the muscles of mastication on both sides became involved. Five convulsive attacks were observed during an examination lasting one hour, and they were all of the same character.

Dr. Hansell examined the eyes and reported: O.D., senile choroidal atrophy; O.S., senile choroidal atrophy; otherwise eyes are in good condition for the age of the patient.

June 22. The man cannot be aroused, and is said to have a convulsion every few minutes.

He was referred to Dr. Edward Martin's service for operation and on June 23 an osteoplastic flap was raised. The upper line was $\frac{1}{2}$ inch to the right of the midline and extended about $2\frac{1}{4}$ inches in front and $1\frac{3}{4}$ inches behind the fissure of Rolando. The base line was about at the upper border of the auricle and extended one inch in front and one inch behind the end of the fissure of Rolando. The portion of the brain at the anterior part of the opening was paler and palpation seemed to Dr. Martin to show that this portion was not so dense as that more posteriorly, and it was suggestive of a subcortical lesion such as a cyst. This area when explored appeared to be somewhat softened. A small piece of this apparently softened tissue was removed for microscopic study.

The convulsions confined to the left side continued after the operation, beginning within a few hours. From 8.45 P.M., June 23, to 7 A.M., June 24, the patient had forty-one convulsions confined to the left side. The convulsions occurred frequently during the few days preceding death, and the patient lay in the position of pleurosthotonos, with the body drawn to the left and the eyes partially drawn to the left even during the intermissions between the convulsions. In the convulsions the abdominal muscles on the left side were

in tonic contraction. The man became gradually weaker and died June 25, 1904, the convulsions continuing until death.

Only a portion of the right hemisphere was obtained at the necropsy. After this hemisphere had been hardened in 10 per cent. formalin, horizontal sections were made through it. The portion which seemed soft at the operation was found to be the seat of a small subcortical tumor, not reaching quite to the surface of the brain (Figs. 1 and 2). This tumor measured 1.5 cm. by 1.5 cm. by 0.5 cm. from above downward. It was situated in the posterior end of the second frontal convolution and did not extend into the precentral convolution. Microscopic examination showed it to be a glioma.

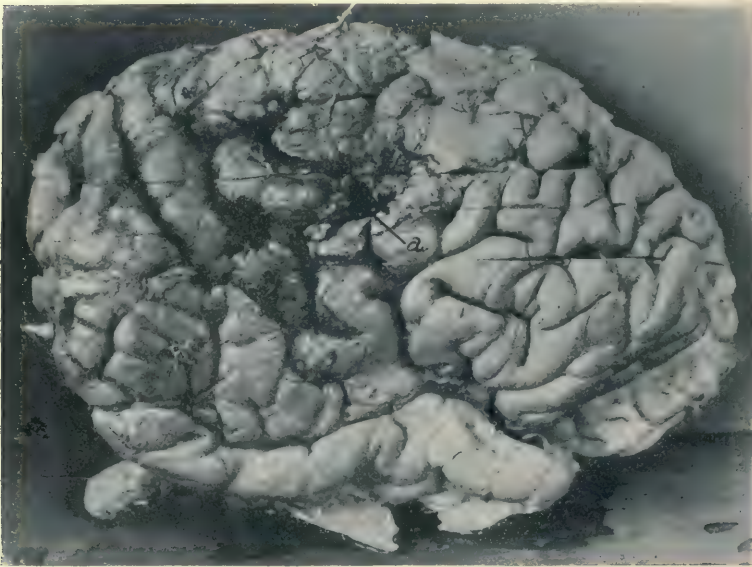


FIG. 1.—Photograph of a portion of the right cerebral hemisphere. The line *a* indicates the position of the tumor and the level of the horizontal sections shown in Fig. 2.

The continuance of the convulsions, notwithstanding the exposure of the brain by an osteoplastic flap and relief thereby of intracranial pressure, was suggestive of an intracranial lesion causing irritation. In a case of mine of idiopathic epilepsy with convulsions of the Jacksonian type, recently operated upon by Dr. C. H. Frazier, the exposure of the brain caused a marked arrest of the convulsions.

The implication in the convulsions of the muscles on both sides

innervated by the facial nerve, only in the upper part of the face, and of the muscles of mastication on both sides is interesting, and is probably to be explained by the more nearly equal innervation from both sides of the brain of those muscles on each side of the body that commonly functionate together. This bilateral implication of the muscles of the upper part of the face in convulsions otherwise Jacksonian in type I have observed repeatedly. Interesting also was the deviation of the eyes to the left during convulsions of the left side of the body. In reporting a similar case



FIG. 2.—Horizontal sections showing the tumor (a) in situ.

in 1902 I remarked that: "This clinical observation supports the view that the chief centre for conjugate deviation must be near the motor centres of the limbs, because if these centres were widely separated we should not expect in a case like this, in which the convulsions were confined to one side of the body, to find conjugate deviation of the head and eyes during the convulsions."¹ The present case is valuable because the movements of the eyeballs

¹Journal of Nervous and Mental Disease, 1903, p. 161.

to the left occurred only during the convulsions and the tumor was in the posterior part of the second frontal convolution, the portion of the brain believed to be the chief centre for lateral movements of the eyeballs. I have recently, however, had a case in which more persistent conjugate movements of the eyeballs were produced by a hemorrhage in the parietoöccipital region.

The most interesting feature of this case is the duration of the Jacksonian convulsions during eight years, and resulting from so small a tumor; for it is presumable that the tumor was the cause of the convulsions. We are inclined to give a grave prognosis when a diagnosis of cerebral tumor is made, and properly so, but it is well to remember that occasionally a tumor may grow very slowly, especially if it is a glioma, and may still be small after several years.

A glioma may grow to large size without causing symptoms. Thus in a case reported by Rosenblath,¹ a large, partially calcified glioma of the left frontal lobe was found, but symptoms are said to have been present only about five weeks and to have developed after a fall. In another case the symptoms of the glioma had been present only three or four weeks. As Rosenblath says, there are more cases reported of glioma with a long course than with a short one, but those that have existed for years, even decennaries, are such as appear macroscopically as hypertrophied parts of the brain. The soft tumors, rich in cells, are of more rapid growth.

I have seen recently in consultation with Dr. J. W. McConnell a case in which a large glioma of the right frontal lobe had caused symptoms only during the six weeks preceding death. This case will be reported by Dr. McConnell.

The occasional long duration of brain tumors has been discussed recently by H. H. Hoppe.² He says that some cases with unusually long remissions of the symptoms, like Osler's case, finally end fatally and he refers to the cases reported by Oppenheim, Gowers, Bernhardt, and Russell.

"Psammoma, lipoma, and cholesteatoma may attain a certain

¹ Deutsche Zeitschrift f. Nervenheilk., vol. xxxi, Nrs. 5 und 6, p. 342.

² Journal of Nervous and Mental Disease, February, 1907.

size and then cease to grow. Cysticerci and echinococci may die and shrink. The observations confirmed by autopsy are exceedingly rare. Nonne was able to find but four cases, those of Bruns, (Oppenheim, and Simeon.

"Aneurysms may cease to grow or become obliterated by the formation of a clot and the brain accommodates itself to the pressure (Oppenheim, Hutchinson, Hodgson, and Humble). Solitary tubercles may undergo caseation, calcification, and then become encapsulated, causing no further damage (Wernicke, Starr, Knapp, Gowers, Babinski, and Sahlberg). These observations are confirmed by the pathological observations of Simeon, who reports an autopsy in which a calcified tubercle was found which must have been in the brain for thirty years. Kirschberger's case shows how a tubercle may cause the signs of a brain tumor for a year and a half and the patient recover and remain well for six and one-half years. The autopsy in this case showed that the tubercle had become calcified. Gumma may become absorbed and disappear." [To my mind the absorption of a macroscopic gumma is doubtful.]

"Long remissions and even permanent disappearance of symptoms may occur in brain tumor, as numerous cases on record show. These cases, however, would hardly come under the head of pseudotumors with a complete disappearance of symptoms, because they had been regarded during life as cases of epilepsy of many years' duration, and after death partly or completely ossified tumors had been found as the cause of the epilepsy."

Hoppe thinks that a case he has observed shows that a pathological mass may be present in the cortex of the brain for forty years and cause no interference with the ordinary daily life of an individual. A man, aged sixty years, died in his service at the Cincinnati Hospital from acute meningitis. He stated that at the age of twenty-one his left arm became paralyzed, but that it soon recovered. He had lived the life of a farmer and teamster for the past forty years. With the exception of some weakness of the left arm and a constant headache he had always been well.

The autopsy revealed a hard, indurated, cartilaginous mass about the size and shape of a peanut, situated in the cortex, at

the posterior end of the first frontal convolution of the right side, and pressing upon the ascending frontal convolution. This tumor was partly cortical and partly subcortical, was not firmly adherent, and was surrounded by recent softening. On splitting it open a very hard, stony, calcareous centre, with a hard, cartilaginous covering was found.

The paralysis of motion and disturbance of sensation of the left limb in my patient were probably the result of exhaustion from the convulsions, and these symptoms became more intense as the spasms continued. Death was probably caused by exhaustion. It is remarkable that the motor cortex was not invaded.

Even when the brain was exposed at the operation we could not be sure of the existence of a tumor, although we found an area which appeared to be pathological and was the diseased cortex over the tumor. It did not seem advisable to excise this, as we could not be certain of the nature of the lesion, but from the later developments it is much to be regretted that the excision was not performed.

Where Jacksonian convulsions are persistent, even though there may be no other symptom, it seems advisable to operate. We probably run less risk by operating than we do by permitting the convulsions to continue without surgical intervention, although we should remember that occasionally status hemiepilepticus may occur without organic lesion, as emphasized especially by Leo Müller,¹ Nonne,² and Bernhardt.³ In another case in which Jacksonian convulsions were the only symptoms I recommended operation and a small tumor was found and excised. It is in cases of cerebral tumor such as these that we may hope the most from surgery, and probably only when a glioma is so small are we able to fully remove it. I am indebted to Dr. Alfred Reginald Allen for the photographs.

¹ Deutsch. Zeit. f. Nervenheilk., vol. xxviii.

² Neurol. Centralb., 1906, Nr. 12, p. 589.

³ Berl. klin. Woch., November 5, 1906, p. 1443.

REMARKS BY DR. MARTIN.

At the time this patient presented himself to the surgical ward for operation his condition was such as to justify no good from a conservative treatment and very little from operation. Since, however, localizing symptoms were persistent and very characteristic, since the exposure of the brain can be accomplished rapidly and with little shock or hemorrhage, an osteoplastic flap laid out by Dr. Spiller in suchwise as to give free access to the Rolandic area was rapidly turned back. Bleeding from the scalp wound was obviated by an elastic ligature passed around the skull from the forehead to the occiput and secured from slipping by three catgut sutures. The electric osteotome failed to work because of the burning of the fuse. By means of four trephine openings and a powerful set of rongeur forceps the bone was turned back almost as rapidly as would have been the case with the saw and cutting drill. The dura appeared normal and was pulsating. It was turned back, exposing a soft, bleached area merging with the healthy surrounding brain by imperceptible gradations. A portion of this was excised for microscopic examination. The dura was sutured and the osteoplastic flap brought back to position. There was very little bleeding and no appreciable shock as the result of this operation and the wound ran a reactionless course.

The matter to be regretted is that the operation was incomplete. The neurological localization was perfect. The seat of the lesion was regioned, but because of the lack of definite surface findings and the unfavorable condition of the patient a thorough subcortical exploration was not made. Such an exploration was certainly justified by the symptoms, and in this particular instance it might have resulted in a cure.

THE CLINICAL RESEMBLANCE OF CEREBROSPINAL SYPHILIS TO DISSEMINATED SCLEROSIS.¹

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THE diagnosis between cerebrospinal syphilis and multiple sclerosis, while not usually regarded as difficult, may in some cases be very perplexing. Both multiple sclerosis and syphilis vary in their symptomatology according to the location of the lesions, so that difficulties in clinical diagnosis might be expected. In this paper no special reference is made to those cases of disseminated syphilitic lesions which resemble multiple sclerosis pathologically as well as clinically, nor is any possible causal relationship between the two diseases discussed.

A case in which a positive diagnosis between cerebrospinal syphilis and multiple sclerosis for a long time seemed impossible was under the care of one of us (Dr. Spiller) during several of his terms of service in the Philadelphia General Hospital, and was frequently presented in lectures to the students as probably one of multiple sclerosis. The patient died two weeks after the termination of one of these terms of service, and we are, therefore, indebted to Dr. James Hendrie Lloyd for the pathological material.

P. B., aged twenty-eight years, was admitted to the Philadelphia General Hospital November 10, 1902, complaining of tremor, difficult locomotion, and a steady, constant pain in the legs just above the knees.

His father was living and well; his mother was dead of an unknown cause. Three brothers and one sister were living and well. The patient was married and the father of one healthy child. He ad-

¹ Read at a meeting of the Philadelphia Neurological Society, January 22, 1907.

mitted having had gonorrhœa when fifteen years of age and chancroid when about twenty years of age, but denied any history of syphilitic infection; subsequently, a few months before his death, he admitted having had syphilis. He denied excessive use of alcohol, and was always apparently strong and healthy. About six months previous to the onset of his present trouble he had his hands and feet frost-bitten while shovelling snow, but they were not much swollen, and he worked the next day as usual.

His present trouble began about two months before his admission to the hospital in 1902, when he noticed that when walking he would stagger and have pains in his legs. The pains were slight and not sharp or shooting in character. Three weeks after the legs were affected he noticed a beginning affection of the hands and arms, the nature of which is not recorded, unassociated with any pain, however. He stopped work about one week previous to his admission to the hospital, on account of repeated falls from his sweeping machine.

On his admission to the hospital it was noted that his gait was distinctly ataxic and that his station was very poor, so that he fell when he attempted to take the position of "attention" with his eyes open or closed. His irides reacted normally to light and in accommodation. He had general tremors of the limbs. His speech was affected and his tongue was tremulous. The knee-jerks were absent. Ankle clonus was slightly manifested and plantar irritation showed a slight extension of the great toe on the left side. The examination of the chest and abdomen was negative.

October 21, 1903. An ocular examination was made by Dr. E. A. Shumway; the ocular movements and the pupillary reactions were normal. Vision, O. D. $\frac{5}{20}$, O. S. $\frac{5}{15}$. The ophthalmoscope showed a pallor of the temporal half of the right optic nerve and a decided simple atrophy of the left optic nerve. The visual fields of both eyes were concentrically contracted, but especially that of the left.

November 12th. It was noted that the patient was exceedingly nervous and irritable. He did not have nystagmus. The upper extremities showed a coarse tremor on movement and he was unable to button his clothing. He had slight loss of voluntary power, the dynamometer registering 50 in each hand. With his eyes shut, he recognized correctly familiar objects placed in his hands. The tendon reflexes were apparently lost in the upper extremities, but the tremor made the examination difficult. His gait was ataxic, and he could not stand unsupported with his eyes open or closed. The knee-jerks were lost. Ankle clonus was present on each side, but the Babinski sign was apparently present only on the left side. He had no loss of sensation.

February, 1905. It was noted that the knee-jerks and ankle clonus were absent on both sides; that there was a marked intention tremor of the hands and a staccato speech.

February 20. An ocular examination was made by Dr. Charles A. Oliver. The right pupil was larger than the left. Both irides were sluggish to light, but prompt in accommodation and convergence. The movements of the external ocular muscles were good in all directions. The papulomacular wedge and the tissues in the optic nerve, downward and outward, were partially atrophic. The left optic nerve was more diffusely atrophied than its fellow. Efforts to outline the visual fields were unavailing owing to the patient's mental condition.

March 10, 1906. Notes made by Dr. Wm. G. Spiller. His condition was much as it had been for several years, only the man was much weaker than he had formerly been. He had difficulty in showing his teeth, and when he attempted to do so there were irregular movements about his mouth of the character of an intention tremor. He wrinkled his forehead well on each side, but was unable to draw up either corner of his mouth separately. When he closed his eyes there was a tremor of the muscles above the eyelids. This intention tremor of the muscles of the face was very striking and was a new sign. The movements of the eyeballs were free and there was no nystagmus. Sensation for touch and pain was normal in the face. The tongue jerked violently backward and forward when he attempted to protrude it, but when at rest it presented no tremor. The tremor of the tongue was an intention tremor. The masseter muscles contracted well. Speech was very difficult, tremulous, and scanning. The upper limbs were not wasted and showed some small white scars, which the patient now confessed were caused by syphilis. The slightest voluntary movement of either upper limb produced marked intention tremor. Every few minutes he had involuntary jerkings of one or the other upper limb. The biceps tendon reflex and the triceps tendon reflex were lost on each side. Sensation to touch and pain seemed to be unaltered in the upper limbs. There was no wasting of the hands or of the trunk muscles. The grasp of each hand was feeble. The lower limbs were not muscular, but there was no localized atrophy. The patellar reflex was completely lost on each side, even on reinforcement. The lower limbs could be moved in all parts, but the movements were feeble and intention tremor was pronounced in most of them. The tendo Achillis jerk was lost on each side. Babinski's sign was not distinctly present on either side, the movements of the toes being uncertain. Sensation for pain was normal in the lower limbs, but his answers regarding tactile sensation were unreliable because of his mental dulness. If not aroused he had a tendency to fall into a slightly stuporous state.

March 23. Mental aberration was present; he was delusional and swore considerable.

April 10th. He was somewhat quieter. Pulse rapid, but of good tension; temperature 101°; respirations increased in frequency

and shallow. Examination of the chest was negative. He died on April 16, at 8 P.M.

At necropsy the brain and spinal cord appeared normal. They were removed and preserved in 10 per cent. formalin solution for histological examination.

In sections from the lumbar region of the spinal cord stained by the Weigert hematoxylin method, the posterior columns appeared much degenerated in the manner usually seen in tabes, the cornu-commissural zone and the oval field of Flechsig escaping. The lateral columns in the region of the crossed pyramidal tracts were a little less deeply stained than the rest of the anterolateral columns. Sections stained by acid fuchsin and hemalum showed round-cell infiltration of considerable intensity in the pia, but not about the bloodvessels within the spinal cord. The bloodvessels in the pia and those in the anterior roots were much congested and thickened. The nerve cells in the anterior horns stained by the thionin method were in a fairly good state of preservation, but here and there a very highly diseased cell was found.

The round-cell infiltration of the pia was more pronounced in the thoracic region than in the lumbar cord, and there was also a slight cellular infiltration about some of the bloodvessels of the spinal cord, especially in the sclerotic areas in the posterior columns. The posterior columns, except the cornu-commissural zones and along a part of the posterior septum, appear intensely degenerated in sections stained by the Weigert hematoxylin method. The crossed pyramidal tracts were a little less deeply stained than the rest of the lateral columns. The columns of Lissauer were degenerated.

In the cervical region the posterior columns were still intensely degenerated, and the degeneration resembled that of tabes. The lateral columns appeared to be normal. Round-cell infiltration of the pia was about as intense as in the lumbar region, but there was no distinct infiltration about the bloodvessels in the spinal cord. The nerve cells of the anterior horns were in about the same condition as those in the lumbar region; here and there a nerve cell was seen that was much degenerated.

Sections of the medulla oblongata stained by the Weigert hematoxylin method appeared to be normal. The hemalum-acid-fuchsin stain showed the round-cell infiltration of the pia to be as intense here as in the spinal cord, but it was not present about the bloodvessels within the nervous substance. Sections through the pons and through the cerebral peduncles showed the same round-cell infiltration in the pia, but the sections appeared otherwise normal. The cells of the nuclei of the hypoglossal, facial, abducens, trochlear, and oculomotor nerves, as well as the nucleus ambiguus, appeared normal when stained by the thionin method.

The pia of the right and left paracentral lobules was infiltrated with round cells and there was a distinct round-cell infiltration about

the bloodvessels within the cortex. The cells of Betz appeared to be in a very fair condition, although here and there a cell was found partly degenerated.

Round-cell infiltration about the optic chiasm was only of moderate intensity, and was also found within the chiasm about the bloodvessels. The optic nerves and chiasm stained well by the Weigert hematoxylin method.

Sections of the cerebellum showed a moderate round-cell infiltration of the pia mater. Some of the Purkinje cells showed peripheral displacement of the nucleus.

The oculomotor nerves appeared to be partially degenerated and presented considerable round-cell infiltration within the nerves and an overgrowth of the connective tissue. Both sixth nerves appeared partially degenerated, when stained by the Weigert hematoxylin method, and there was considerable overgrowth of connective tissue within them. The motor and sensory roots of the left fifth, the left seventh, the left ninth and tenth, and the right twelfth nerves appeared fairly normal by the hemalum-acid-fuchsin and Weigert hematoxylin stains.

In order to show the resemblance of this case to one of multiple sclerosis it may be well to give a brief summary of it.

A young man positively denying, during several years, syphilitic infection, presented marked ataxia of gait, intention tremor of the limbs, and, a month or two before death, of the muscles of the face; scanning speech; at first normal iridic reactions, later Argyll-Robertson pupils, with unequal pupils; pallor of the temporal side of the right optic nerve and simple atrophy of the left optic nerve; and vertigo. Pain was never a prominent symptom. Remissions did not occur during the years he was in the hospital. Nystagmus was not observed. Shortly before his death, when some scars on his limbs were discovered and a history of syphilis obtained, the diagnosis of cerebrospinal syphilis was entertained, although it was not forgotten that a patient with multiple sclerosis is not exempt from syphilis.

The lesions found were those of meningo-encephalomyelitis, consisting chiefly of round-cell infiltration and degeneration of the posterior columns of the cord.

The patient was married and was the father of a healthy child. He positively denied having had syphilis until a short time before his death; on the other hand, his occupation, in which he was constantly exposed to the weather, and a history of having had a severe exposure to cold a short time before the onset of his illness, furnished a possible etiology for multiple sclerosis. The influence of exposure to cold and damp in the causation of multiple sclerosis has been referred to by Krafft-Ebing, Spiller, and others. Pain was never a marked feature of the case, and at no time was of the sharp,

shooting character usually associated with inflammatory lesions of the meninges involving the posterior roots. The attacks of vertigo, as indicated by the repeated falls from his sweeping machine, are also in favor of the diagnosis of multiple sclerosis.

The gait in many cases of spinal syphilis, as also in multiple sclerosis, is of the spastic-paretic type, but the ataxic type is common. In this case there was marked ataxia in gait and station, which, however, was not increased by closing the eyes, so that it resembled the incoördination of gait characteristic of multiple sclerosis. The tremor was of the so-called intention type, was widely distributed, and involved, late, the tongue and the muscles of the face as well as the limbs. This intention tremor is regarded generally as being diagnostic of multiple sclerosis. The speech was "scanning," but there was no nystagmus. Uhthoff² found nystagmus present in only 58 per cent. of cases of multiple sclerosis, so that its absence in this case was not regarded as particularly significant. In six cases observed by E. W. Taylor,³ in which the diagnosis was confirmed by necropsy, nystagmus seems to have been observed in only one or two, and then only on lateral deviation of the eyeballs. Uhthoff calls attention to the analogies between the ocular changes that occur in cerebrospinal syphilis and those of multiple sclerosis; the frequency of visual changes without ophthalmoscopic findings and recovery from them, the unilaterality of the symptoms, with the great infrequency of double total blindness. The changes in multiple sclerosis are of a less severe grade than those of syphilis; hemianopsia is rare; ophthalmoscopic changes are less marked and may last longer without alteration of vision. Optic neuritis is relatively seldom found in multiple sclerosis. A simple atrophy and pallor occur in both diseases. Parinaud⁴ calls attention to the fact that paresis of a single eye muscle ending in recovery is frequent in multiple sclerosis, while complete or lasting paralysis rarely occurs, which it does, however, in syphilis. The findings in our case of a simple atrophy of one optic nerve and an atrophy limited to the temporal half of the other were very suggestive of multiple sclerosis.

The knee-jerks and the Achilles jerks are usually exaggerated in both multiple sclerosis and cerebrospinal syphilis, but this symptom depends on the location of the foci of the disease, and as has been shown by one of us (Spiller⁵), the knee-jerks may be lost in multiple sclerosis if the sclerotic foci invade the reflex arcs.

Summing up the symptoms in this case, it is evident that there was enough or more than enough to make the diagnosis of multiple sclerosis, and that there were no symptoms that were incompatible with such a diagnosis. Paoli Pini,⁶ reporting cases from Oppen-

² Archiv f. Ophth., Band xxxix, Heft 1 und 3.

³ Journal of Nervous and Mental Disease, June, 1906.

⁴ Progrès méd., August, 1884.

⁵ AMER. JOUR. MED. SCI., January, 1903.

⁶ Deut. Ztschr. f. Nervenheilk., vol. xxiii, p. 267.

heim's clinic, does not hesitate to make the diagnosis of multiple sclerosis on far less evidence. The literature contains a number of papers devoted to the resemblance of cerebrospinal syphilis to multiple sclerosis, but most of the reported cases are without confirmation by necropsy. B. Sachs⁷ pointed out the close resemblance between multiple sclerosis and cerebrospinal syphilis, and remarked that spastic paraplegia of the lower extremities, ocular palsies, optic-nerve atrophy, loss of pupillary reflexes, difficulties of speech, mental changes, apoplecticiform seizures, marked remissions in all the symptoms are common to both affections. Nystagmus, intention tremor, and scanning speech are characteristic of multiple sclerosis, and are supposed to be absent in syphilis of the nervous system, but he has seen nystagmus and peculiarities of articulation resembling scanning speech in syphilis. In some cases of syphilis a tremor occurs which differs from the ordinary intention tremor only in that it is persistent and is aggravated on voluntary movements.

Sachs emphasizes the importance of the iridic reflex in making a diagnosis between multiple sclerosis and cerebrospinal syphilis, and considers the complete immobility of one or both pupils strong evidence in favor of syphilis rather than of multiple sclerosis. Indeed, in one of Cassirer's cases of doubtful diagnosis Sachs says that he (Sachs) did not regard the case as one of syphilis because the pupillary reflexes remained normal. In our case of syphilis pupillary rigidity was not present; at first the reflexes were normal; two years later the Argyll-Robertson pupil was observed. Nonne,⁸ in his monograph on syphilis of the nervous system, says, in regard to the relation of syphilis to multiple sclerosis, that the latter cannot be mistaken for the former when the clinical picture of multiple sclerosis, as drawn by Charcot, is considered. The course of syphilis of the nervous system is different, and scanning speech, rotatory nystagmus, intention tremor, the general tremor of the trunk and head are not observed in spinal syphilis; this statement can no longer be accepted. If a patient with multiple sclerosis has a history of syphilitic infection the latter proves nothing, and it is probable that syphilis plays no part in the etiology of multiple sclerosis.

Oppenheim⁹ thinks that the symptoms of cerebrospinal syphilis may closely resemble those of multiple sclerosis, especially in cases with spastic paraplegia. In both diseases there are cerebral and spinal symptoms, and in each remissions and exacerbations occur. Disturbance of vision, anesthesia, paresthesia, etc., may vary in intensity in each. However, it is rare for the diagnosis to be difficult for a long period, and yet it was difficult in our case for four years. The cerebral symptoms of multiple sclerosis, he says, are very different, viz., nystagmus and scanning speech; and the tremor is almost

⁷ New York Med. Jour., 1891; Phila. Med. Jour., 1898.

⁸ Syphilis und Nervensystem, S. Karger, Berlin, 1902, p. 306.

⁹ Die syphilitischen Erkrankungen des Gehirns, 1903, 2d ed., p. 5.

pathognomonic; stupor does not occur in multiple sclerosis except in the rare apoplectiform attacks, and is almost always observed at some period or other in cerebrospinal syphilis; but it was not present in our patient until a month or two before death. Oppenheim confesses that he has not always been able to decide whether a certain case was one of cerebrospinal syphilis or multiple sclerosis. The partial optic atrophy is the typical form of disease of the optic nerve in multiple sclerosis; when it occurs in syphilis it is usually accompanied by much impairment of sight, whereas an equal degree of atrophy in multiple sclerosis impairs the sight very little. In our patient, in 1903, vision was $\frac{5}{20}$ in the right eye and $\frac{5}{15}$ in the left. Pupillary symptoms are rare in multiple sclerosis.

The spinal symptoms may be very similar in the two diseases. It is strange, he says, that authors have paid so little attention to the differential diagnosis between cerebrospinal syphilis and multiple sclerosis. He refers, however, to the writings of Sachs, Sanger, Gilbert and Lion, Schuster, Unthoff, Reinhold, Sottas, Marie, v. Bechterew, Cassirer, and Pini, and to these should be added those of Gowers and Krewer.

Some authors hold that syphilis may cause areas of disseminated sclerosis, but does not produce exactly the same symptom-complex as does the true multiple sclerosis. This subject is discussed recently by Guinio Catola.¹⁰ He refers to a case reported by Schuster,¹¹ in which symptoms of disseminated sclerosis were associated with syphilitic ulceration of the pharynx, indurated glands, and periostitis of the right tibia. All symptoms disappeared under anti-syphilitic treatment. This case probably was similar to the one we describe, as one could hardly expect sclerotic areas to disappear under any treatment, and it shows in association with our case that the symptoms of disseminated sclerosis may be caused by the syphilitic infiltration and vascular lesions. The symptoms in Schuster's case were intention tremor of the upper and lower limbs, dragging of the feet, exaggeration of the tendon reflexes, slight contractures of the limbs, and true nystagmus. Catola reports two cases with necropsy which he thinks bear on the question of the syphilitic origin of multiple sclerosis. Müller,¹² in his recent book on multiple sclerosis, refers to the resemblance this disease may have to cerebrospinal syphilis, but draws his conclusions from the literature rather than from personal experience.

The difficulties in the diagnosis have been regarded as the cause of the apparent comparative rarity of multiple sclerosis in America. It may be that many cases, especially those of the *formes frustes*, are overlooked by superficial examination, but, on the other hand, there seems at present a danger that much will be called multiple sclerosis that in reality is some other disease.

¹⁰ Nouvelle Iconographie de la Salpêtrière, July and August, 1906, p. 337.

¹¹ Berl. klin. Woch., 1885.

¹² Die multiple Sklerose des Gehirns und Rückenmarks, p. 250.

**SENSORY AND MOTOR DISTURBANCES IN PARTS ABOVE
THE DISTRIBUTION INVOLVED BY DEFINITE ORGANIC
LESIONS OF THE SPINAL CORD.***

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The object of this paper is to call attention, first, to the gradual extension of the areas of disturbed sensation above the peripheral distribution related to definite lesions of the spinal cord, and secondly, to the marked increase in the tendon and skin reflexes in the same parts, and in parts above. These facts have not, heretofore, been noted.

These observations are based upon three cases, one with necropsy. In all, the lesions were of long standing, being 35, 29 and 4 years in duration, and the sensory phenomena were carefully studied and noted at varying intervals. The lesions were of traumatic origin, and the symptoms were of such clearness that it was possible in each case to make a clear-cut diagnosis in definite localities of the spinal cord.

The cases are as follows:

Case 1. Morgan. Was admitted to the nervous wards of the Philadelphia Hospital June 13, 1899, where he still is. His past and family history are of no importance, has never used alcohol or tobacco; and has never had syphilis.

When 27 years of age he was struck across the lumbar spine by a piece of iron and rendered unconscious. When he recovered, he was unable to use his lower limbs, and has never had more than slight use of them since. He has never had any bladder or rectal disturbances, or bed sores. Considerable pain was present in his lower limbs for some years after the injury, and girdle sense has always been present.

Notes by Dr. Van Epps made in January, 1900, state that the patient had from January 1 to 14 herpetic eruptions accompanied by much pain in the left side of the body extending from mid to mid line laterally and from costal margin to iliac crest posteriorly, and the pubic line anteriorly. The vesicles gradually disappeared but the pain continued for some time.

Examination on November, 1900, 28 years after the injury,

*Read at the meeting of the American Neurological Association, May 7, 8 and 9, 1907.

by me, denoted almost total loss of power in the lower limbs. They were considerably wasted and spastic, more so the right. The cremasteric, patellar and plantar reflexes were present and prompter than normal; ankle clonus was present on each side, but the Babinski reflex could not be obtained.

Sensation for all forms was absent to a line corresponding



Fig. 1.



Fig. 2.

Fig. 1. The shaded areas represent disturbed sensation in 1900.

Fig. 2. The shaded areas represent disturbed sensation in 1907.

with the umbilicus. Above this point pin prick was appreciated, but touch and temperature sensations were not normally recognized until two inches above the umbilical line; just above this line there was a zone of hyperesthesia and the patient complained here of a girdle sense.

Examination by me on February, 1907, seven years after the last examination and thirty-five years after the injury,

showed practically the same condition as to his motor power in the lower limbs. He could only slightly move the toes and knees. The cremasteric and patellar reflexes were still increased, but ankle clonus could not be obtained; the Achilles jerks were present. Plantar irritation produced flexion of all the toes. The bladder and rectum were normal.

The state of the sensation altered considerably. In my first examination of this year, he was unable to recognize touch, pain and temperature sensations over the lower limbs and abdomen up to the line of the nipples, and sensation did not really become normally appreciated until about the first intercostal space. Subsequent examinations have confirmed this, with the exception that pin prick was occasionally interpreted correctly below the nipple, but never more than two inches above the line of the umbilicus. (Fig. 2.) Bone sensation as tested by the tuning fork was diminished in the lower ribs, but was gradually better appreciated as the upper ribs were approached, and was normal about the second rib.

Power in the upper limbs was normal, and there was no apparent atrophy present. In the finger to nose test some ataxia was noticeable, but this was not marked. There was some hypotonia in the elbow joints. The sense of position was about normal, but on one occasion when absolute relaxation was obtained, it was demonstrated to some extent in the fingers, wrist and elbow joints. The biceps, triceps and wrist reflexes were prompter than normal, more so in the right side. On tapping with the percussion hammer over the chest and shoulder prompt contraction could be obtained in the corresponding muscles, so that practically reflexes could be obtained wherever desired.

X-ray examination made by Dr. Leonard D. Frescoln showed an inflammatory thickening (probably periostitis) around 10th to 12th thoracic and 1st lumbar vertebræ.

Summary. A man of 27 years, as a result of a severe blow to the 10th, 11th and 12th thoracic and first lumbar vertebræ sustained complete paralysis of the lower limbs. There must have been an almost complete transverse myelitis of the 9th, 10th, 11th and 12th thoracic spinal segments. An examination made by me 28 years after the injury denoted a spastic paralysis of the lower limbs with increased tendon reflexes, and disturbed sensation up to a line 2 inches above the umbilicus. The bladder and rectal functions were at no time involved. Examination made also by me 7 years later and 35 years after the injury showed the same condition as to motor power in his lower limbs, but sensation was disturbed to the first intercostal space. The tendon and skin reflexes in the upper limbs and in the reflex arcs of the chest and shoulder muscles were markedly exaggerated.

Case 2. Egan. Was admitted to the nervous wards of the Philadelphia General Hospital, May 11, 1892. His family and past history were of no importance and there was no history of alcoholism or syphilis. In 1884, at the age of 35, the patient lost his balance while on a running freight car, and fell to the track, striking his lumbar spine. He then lost

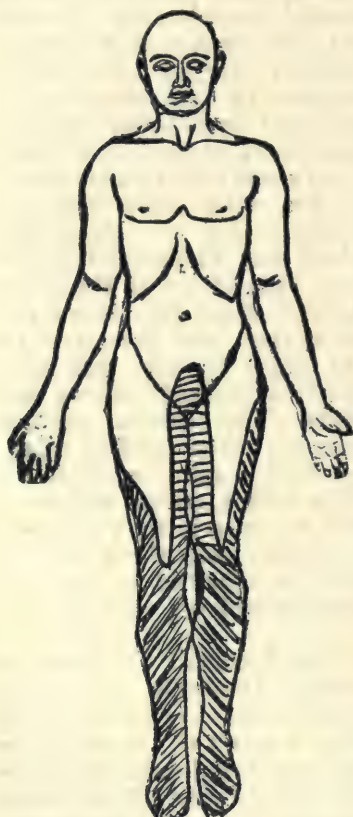


Fig. 3.



Fig. 4.

Fig. 3. The shaded areas represent disturbed sensation in 1899.

Fig. 4. The shaded areas represent disturbed sensation in 1907.

almost all power in his lower limbs and control of his bladder and rectum.

Notes made in June, 1899, 15 years after the injury, by Dr. G. E. Pfahler, state that the patient had loss of power in the lower limbs with the exception of some ability in flexing the right thigh on the abdomen. Bladder and rectal functions were completely lost. Sensation was lost as indicated in chart No. 3.

Examination by me in February of this year (1907), 23 years after the injury and eight years since the last examination, showed almost the same condition as regards the motor power of the lower limbs. The only movement possible was that of slight flexion of the right thigh on the abdomen. The toes were extremely hyperflexed and cyanotic. All of the tendon reflexes in the lower limbs were lost. Plantar irritation produced no movement of the toes. Bladder, rectal and sexual functions were totally lost.

Sensation for all forms was totally lost in the lower limbs and buttocks, with exception of a small area just in the inner surface of both thighs, and over the lower parts of the abdomen, to an irregular line about two inches below the umbilicus. From this line to a line drawn about 1 to 1½ inches above the umbilicus, sensation became gradually better appreciated, being normal at the above point. (Chart No. 4.)

Power and movement was normal in the upper limbs. The biceps and triceps tendon reflexes were very prompt, more so on the left side. On tapping with a percussion hammer over the chest and shoulders prompt contraction could be obtained in the corresponding reflex arcs. The abdominal reflexes were lost on both sides, as were also the cremasteric. In the finger to nose test, some ataxia was present, more so in the left limb. Sensation was normal in the upper limbs.

The X-ray examination denoted caries of the body of the third lumbar vertebra, especially towards the left side.

Summary. A man of 35, as a result of a fall, became completely paralyzed in the lower limbs with total loss of bladder and rectal functions. There must have been a complete transverse myelitis of all portions of the cord below the 2nd and 3rd lumbar segments. An examination made 15 years after the injury denoted almost a complete flaccid paralysis of the lower limbs with the exception of a slight ability to flex the right thigh on the abdomen. The bladder and rectal functions as well as all of the tendon reflexes were totally lost. Sensation was disturbed over both lower limbs, buttocks and perineum with the exception of a triangular area over the front of the thighs. An examination made 8 years later and 23 years after the injury showed the same condition as to the motor power in the lower limbs, but sensation is now disturbed to a line about 1½ inches above the umbilicus. The tendon and skin reflexes in the upper limbs and over the chest and shoulder are markedly increased. There is some ataxia in the upper limbs.

The third case, a brief history of which is only given, does not illustrate the sensory and motor changes which are the subject of this paper, but is used because it was a case of traumatic myelitis in which a careful microscopic examination

was made, and in which the pathological findings are illustrative of the changes which probably have occurred in the two similar clinical cases here recorded.

Case 3. This case was recorded by Dr. Spiller and myself in the *Review of Neurology and Psychiatry*, October, 1904. A young man, because of an injury to the back, sustained complete paralysis of both lower limbs and of the bladder and rectum. All of the tendon reflexes of the lower limbs were lost. When examined by me one year after the injury, sensation was lost to a line corresponding about 2 inches below the umbilicus, and further examination made 3 years later and 4 years after the injury denoted absence of all sensation to a line drawn about 1 inch below the umbilicus.

A careful microscopic examination of sections taken from the upper part of each segment of the spinal cord from the 10th thoracic to the upper part of the cervical enlargement showed that as high as the upper part of the 10th thoracic segment, the cord was completely destroyed. Many normal fibers entered the cord in the posterior roots of the 9th thoracic segment.

Examination of the sections above the 9th thoracic segment stained by the Weigert hematoxylin method showed that the posterior root entrance zones, and the columns of Burdach and Goll were much better and more deeply stained than the other portions of the cord. A marked thickening was also found in the walls of the arteries, besides the usual ascending secondary degenerations.

We have here, therefore, two cases of definite lesions of the spinal cord; one involving the 9th to the 12th thoracic segments; and in the other the spinal cord below the 2nd or 3d lumbar segments. In both the lesions were of long duration, being 35 and 23 years, and the sensory symptoms were studied and noted at intervals of 7 and 8 years respectively. The areas of sensory disturbances increased in the period of 7 years in the first case from a line drawn 2 inches above the umbilicus to the 1st intercostal space, and in the second case from irregular areas over the thighs to a line drawn about 1½ inches above the umbilicus. In these areas of increased sensory phenomena, sensation was lost absolutely only about 2 or 3 inches, and above these parts there was a gradual shading off of disturbed sensation. The important point is that the areas of disturbed sensation have markedly increased above the limits previously found.

What are the causes of this increase in the sensory disturbances? The first answer which naturally presents itself, is an extension of the myelitic process, and secondly, the possible influence of the secondary ascending degenerations.

Let us consider first the possibilities of the occurrence of an ascending myelitis. It is well known that whenever the spinal cord is injured, even in a restricted portion, that the parts both above and below the point of injury may have small areas of hemorrhage or of softening. In fact the whole extent of the spinal cord may be so involved. In most instances after the subsidence of the acute process, these areas of softening and myelitis disappear and no trace of them is to be found, and clinically no symptoms appear. It is more than probable, however, that they are replaced by neuroglial tissue and we may have here the origin of a future sclerotic process.

The spinal cord receives its blood supply from the anterior and posterior spinal arteries, both of which arise from the vertebral, and from the intercostal branches. These intercostals have ascending and descending branches, which anastomose freely with one another and with the anterior and posterior spinal arteries, making an arterial network about the spinal cord, from which numerous small branches penetrate the surface. The gray matter receives its blood supply principally from the anterior spinal arteries, and it is to be noted that this supply is largely from different branches than that of the white matter. It is also to be remembered that the arteries which enter the cord are terminal. Considering this, it can be readily understood how a destruction of a number of spinal segments would cause interference with the blood supply, first of the whole cord, and after the acute symptoms have subsided, especially of the contiguous parts. This arterial obliterative process in common with all degenerations has a tendency to increase, and if the patient lives long enough, would involve considerable portions of the spinal cord.

In the case with necropsy (Case 3) the arterial degeneration is evident. Sections of all parts of the cord show marked endarteritis and this seems to be much more apparent in the lower parts of the cord near the lesion, and especially so in the periphery and in the lateral columns. The vessels in the gray matter seem less sclerosed, the reason for this greater endar-

teritis in the white matter of the cord being the more ready involvement of the arterial supply. This has only recently been called attention to by Allen.¹

It is probable that a similar arteritic process is present in the clinical cases here recorded, but of much greater intensity as the lesions are of 35 and 23 years' duration as compared with 4 years in the case with pathological findings. This endarteritis causes impairment in the nutrition of the parts; this resulting in a gradual necrobiosis and impairment of function. In this is to be found the probable explanation of the increase in the sensory disturbances.

What influence, if any, do the secondary degenerations have? This question has long been in dispute, from the time that Charcot first explained the occurrence of the contractures in hemiplegia as the result of secondary degeneration of the motor columns. Take, for instance, in the case with necropsy. Here as a result of a myelitis in the 9th thoracic segment, marked secondary degenerations are to be found in the cervical cord. In studying such sections stained by the Weigert hematoxylin method it seems as if these extensive degenerations must have some influence, if not upon the neighboring fibers, at least upon the fibers which mingle with them.

Let us try to analyze this. In a lesion of one internal capsule interrupting the motor columns, secondary degeneration results throughout the whole extent of the pyramidal tract. Such degeneration will, at first, only be apparent by the Marchi and later by the Weigert hematoxylin method.

The question is whether this has any influence upon the structures which are associated with it. We assume that there is a connection between the cells of the anterior horns of the spinal cord and the motor columns, and if this is true, as we have every reason to believe, a degeneration of the motor columns should have influence upon these cells, and yet pathologically this has never been satisfactorily demonstrated. That is no argument, however, that this does not actually occur. Clinical evidence shows sufficiently that in hemiplegia there are atrophy and vasomotor disturbance; these have never been satisfactorily explained, but are probably due to a loss of

¹A. R. Allen, University of Pennsylvania Medical Bulletin, 1905.

tone of the whole motor system of which the motor nuclei in the anterior horns of the cord play a prominent part.

When a paralysis occurs, as for instance, because of a capsular lesion, how is loss of power produced? There is as a result of this lesion, immediate disturbance of physiological relation between the cortical motor cells, the pyramidal tracts, the cells of the anterior horns, the anterior spinal roots, the peripheral nerves, and the muscles, tendons and fascia these supply. More than this, there must be some disturbance of the normal relation with the sensory arc.

This disturbance of physiological relation, or "diaschisis," as Von Monakow has termed it, is, of course, immediate, and will become permanent in varying degree; this depending upon the extent of the original lesion. And how is this disturbance of relation manifested? Clinically by loss of power and pathologically by degeneration of fibers. This degeneration, which is called secondary, is progressive as shown by the reaction to the Marchi and Weigert stains. So far our microscopic methods have failed to demonstrate any changes in the cells of the anterior horns, and in the nerves and muscles. This, however, is mainly because of the recuperative power possessed by the motor cells on account of the innervation received from other sources, and the independent trophic action which they possess. We see then that what is termed secondary degeneration is really nothing more than the physiological or mechanical death of a part.

What action, if any, then, has this secondary degeneration or physiological death of a part? Evidently none, for whatever has produced this, has at the same time produced loss of function in the related parts. Secondary degeneration, however, does produce from the very nature of its slow death, some physiological disturbance. Take for instance the motor columns. Even though there is a complete destruction of these as a result of a capsular lesion, other motor fibers, the so-called extra-pyramidal bundles, join the motor columns in their course downwards. Whether these bundles intermingle freely with the motor columns in the pons and medulla, or whether they are independent in their course, has not been fully determined; but it is known that they ultimately join the motor columns in the spinal cord and probably mingle

with them. We can readily imagine that the degenerated or dead fibers must have some detrimental action upon the healthy fibers.

This can be better demonstrated in the so-called ascending or sensory degenerations. Take, for instance, in Case 3, in which because of a lesion in the 9th thoracic segment, the secondary degenerations are marked. As we go higher and higher, newer fibers enter by means of the posterior roots, and join either the posterior columns or the columns of Gowers and the direct cerebellar tracts. These newer fibers intermingle, or at least, accompany the degenerated fibers.

This influence is probably the result of a vascular change. The blood vessels that supply or are in association with a degenerating or degenerated tract must share somewhat in the sclerotic process, and it is conceivable that as a result of this obliterative arteritis, some degeneration is produced in the contiguous or intermingling healthy fibers.

In the case with necropsy a careful examination of sections of the spinal cord above the lesion showed that the fibers in the posterior columns and in the entrance root zones were much better or more deeply stained than the fibers in the other parts. The pallor in the centripetal columns can be explained as the result of secondary degenerations, but this does not explain the apparent degeneration in the motor columns. Retrograde atrophy of the pyramidal tracts has been described, and it occurs in the motor columns above destructive lesions of the spinal cord. This is as we should expect, for when physiological relations are disturbed, as they would be in such case, there must be some disturbance of function in the whole motor system as has been indicated above. This disturbance is manifested pathologically by a downward degeneration of the motor columns below the point of lesion, but is not apparent in most instances in the parts above, at least, by our pathological methods. This is because these fibers still retain their trophic cortical innervation. Reasoning from this, then, retrograde atrophy must occur in every case, in the motor as well as the sensory tracts.

The increase in the sensory phenomena in the parts above the peripheral distribution related to definite lesions of the cord, has an important practical bearing. Most of our obser-

vations upon sensory areas are based on definite lesions of the spinal cord, in which no attention has been paid to a possible increase of the sensory phenomena, as is pointed out in this paper. Manifestly then such observations in which the lesions are of long duration cannot be as accurate as those which are the result of lesions of a shorter time. In this is to be found one explanation of the great variance in the sensory diagrams.

The second part of this paper is to again call attention to the increase of reflexes in the parts above definite lesions of the spinal cord. This clinical fact was first called attention to by me in 1904. ("Clinical Report of Three Cases of Injury to the Lower Spinal Cord and Cauda Equina."²) In this paper I called attention to the increase of patellar jerks in a lesion of the cauda equina. As this subject is an important one and as no other writer has contributed to it, some of the remarks then made pertaining to the subject will be quoted.

"I have been unable to find any reference to this condition in the literature at my command, except in Thorburn's³ contribution to the surgery of the spinal cord. This author mentions the following cases with increase of the patellar reflexes, but pays no special attention to this subject. One case I have found also reported by Franz Volhard.⁴

"First case, cited by Thorburn from Kirchoff; backward crushing of the first lumbar vertebra causing a degeneration of the fourth and fifth sacral segments, the only symptoms being paralysis of the bladder and rectum and increased patellar reflexes. Sexual and sensory changes are not mentioned.

"Second case: a partial compression of the cauda equina about the level of the last lumbar vertebra, causing severe neuralgic pains in the sciatic and pudic distributions, a weakness of some of the muscles of the lower limbs, but no complete paralysis and no anesthesia of the limbs. The bladder and rectal functions were paralyzed. The patellar reflexes were slightly exaggerated, but there was no ankle clonus.

"The third case is cited by Thorburn from Oppenheim, and was one of fracture of the first lumbar vertebra, causing a my-

²The American Journal of the Medical Sciences, May, 1904.

³Philadelphia, Blakiston & Son, 1899.

⁴Deutsche med. Wochens., 1902, No. 33.

elitis of the conus medullaris. There was a slight weakness of the calf muscles, otherwise no loss of power or atrophy. Anesthesia was limited to the peroneal region and buttocks. There was paralysis of bladder and rectum. Here also the knee-jerks were exaggerated, but there was no ankle clonus.

"The fourth case, by Volhard, of a tumor of the cauda equina, as proved by necropsy, with motor and sensory symptoms, absent Achilles jerks, and weakness of the plantar reflexes, but both the patellar reflexes and the cremasteric reflexes were exaggerated.

"In all four of these cases, therefore, the exaggeration of the patellar reflexes was probably caused by lesions below the reflex arc."

In the two clinical cases now recorded, the tendon reflexes in the upper limbs, that is, the biceps and triceps, were very prompt and the skin reflexes over the shoulders and chest were exceedingly exaggerated. Tapping over any portion of the chest, shoulders or back produced prompt and marked contraction in the corresponding reflex arcs. How is this to be explained? Quoting again:

"It is difficult to find a satisfactory explanation for such a phenomenon as this, but evidently the reflex arcs are in some way thrown into a state of excitation in these cases. No writer has made this a subject of careful study, and it seems therefore important to emphasize the fact that a reflex may be increased by a lesion in the spinal cord below the portion in which the reflex arc is represented. Considerable evidence is offered that in the nervous system there are both depressomotor and excitomotor fibers for the different reflexes, these having their origin in the brain; and it may be that other excitomotor fibers arise. We know that spinal roots on entering the spinal cord give off descending branches that pass downward in the posterior columns; the function of these fibers is entirely unknown, but we must assume that in some way they affect the function of the lower segments. It is probable that in a similar manner the lower spinal segments exert some influence over higher segments, and there is no doubt whatever that in the antero-lateral columns degeneration of short fibers occurs upward. It is presumable that these fibers exert some control over higher segments than those in which they

arise, and in this way possibly cause an exaggeration of tendon reflexes."

It is also probable, as has been shown in the present paper, that the disturbance in the arterial supply in the parts above definite lesions of the cord, produces a disturbance in the functions in the motor columns, this being further influenced by the retrograde atrophy. As has been shown by Rothmann, the only constant symptom of a lesion of the motor column is an exaggeration of the reflexes. Here, then, is to be found the explanation of the reflexes in the parts above lesions of the spinal cord.

The following conclusions can be drawn from the study of this paper:

First: An injury to the spinal cord will cause at first interference with the blood supply of the whole cord. If, as a result of such an injury, there should be produced a limited lesion of the spinal cord, there will be interference with the blood supply of the contiguous areas. This interference will be progressive, thus causing arterial obliteration and necrosis in the contiguous parts.

Second: This progressive degeneration will cause disturbance of function, it being manifested clinically by a gradual extension of the areas of disturbed sensation, and by increase in the tendon and skin reflexes.

Third: A lesion in any portion of the spinal cord will cause disturbance of physiological relations in the associated parts. This disturbance is greatest directly after the injury, and becomes less in the course of time.

Fourth: Secondary degenerations "per se" do not produce any direct symptoms, for whatever has produced secondary degenerations, has at the same time caused loss of function in the related parts. Secondary degenerations, however, cause some physiological disturbance. Every degenerating or degenerated tract has healthy fibers from other sources mingling with it. The arterial degeneration present in the involved tract will cause degeneration in these healthy fibers, and of the immediate fibers surrounding the degenerating tract.

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TWO CASES OF THE POLYNEURITIC PSYCHOSIS WITH NECROPSIES AND MICROSCOPICAL FINDINGS.*

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Our main purpose in the present contribution is to give the records, clinical and pathological, of two cases presenting mental symptoms associated with multiple neuritis, with brief reference to a few similar cases in recent medical literature. The outbreak of the multiple neuritis in the two cases here recorded was probably due to a special infection, as in one instance the disease began very late in pregnancy and in the other two or three days after a premature delivery. Both patients were, however, alcoholics.

Soukhanoff and Boutenko¹ and others have indicated that the Korsakoff syndrome might occur in connection with or after various diseases, and, therefore, that the affection is not essentially, or at least not always, an alcoholic one. Korsakoff himself recorded fourteen cases without previous history of alcoholism. Soukhanoff published a case occurring after typhoid fever. He also mentions other cases, some in alcoholic and some in non-alcoholic patients, after perimetritis, jaundice, etc.

In the cases which follow the examination of the cells of the cortex showed that a condition of cells was present due doubtless to neuronal toxemia and strictly comparable pathologically to the affection found in the cells of the spinal cord. If the peripheral affection is held to be a neuritis, there is no reason for not regarding the spinal and cerebral disease as forms of inflammation.

* Read at the sixty-third annual meeting of the American Medico-Psychological Association, Washington, D. C., May 7-10, 1907.

¹ Soukhanoff, Dr. Serge and Boutenko, Dr. Andre, *The Journal of Mental Pathology*, Vol. IV, Nos. 1, 2, 3, 1903.

The observations of Strümpel, those of Cole³ and Sims,⁴ and others, and our investigations in the two cases here recorded are interesting in connection with the fact that one of us (Dr. Mills) in 1886,⁴ some months prior to the publication of Korsakoff's first paper, called attention to the concurrence of spinal and cerebral symptoms in cases of multiple neuritis, holding that these were instances of the concurrence of encephalitis and myelitis with neuritis. The occurrence of psychic symptoms in multiple neuritis was also referred to by him in 1892.⁵ In both of these contributions interesting clinical cases were recorded.

CASE I.—This case is one of three cases of the polyneuritic psychosis recorded clinically by Dr. S. A. Carpenter, interne at the Philadelphia General Hospital, in the sixth volume of the Philadelphia Hospital Reports, published in 1905. She was in the hospital about two and one-half months preceding her death.

M. T., aged thirty-two years, white, was admitted to the Philadelphia General Hospital September 14, 1904. She denied specific disease and had had no miscarriages. She had drunk coffee in large quantities all her life, and for the past six years had been a heavy beer drinker. About six weeks before admission she gave birth to a full term child, which died after five weeks of enteritis.

The present illness dated back to about three months previous to her admission, when she first had a numbness and tingling in her lower extremities from her knees down; shortly after this she noticed that she was gradually becoming weak and that she tired easily. After the first week of her puerperium, on attempting to leave her bed she discovered that she was unable to stand and had pain in her lower limbs. Up to the time of her admission there had been no involvement of the upper limbs. She had been troubled with constipation for two weeks before coming to the hospital, and for the first time was catheterized on the evening of her admission. She said that her sight had been failing for six months before she was admitted.

Examination shortly after admission showed double foot drop; both knee jerks absent and the Babinski not obtainable on either side; an attempt to obtain ankle clonus produced acute pain; both feet were cold and the soles were somewhat moist; sensation to touch and pain was apparently normal.

Her memory for recent events was extremely poor; even though she had been in the hospital only twenty-four hours she was unable to say

³ Cole, Sydney John, *Brain*, Part III, 1902.

⁴ Sims, F. Robertson, *Journal of Nervous and Mental Disease*, Vol. XXXII, No. 3, March, 1905.

⁵ Mills, Chas. K., *The Medical News*, December 18, 1886.

⁶ Mills, Chas. K., *The International Medical Magazine*, February, 1892.

how long it was since she was admitted, and she was never able to remember whether or not she had had her meals, even though she had eaten a few hours before. Two days after her admission, on being asked whether her bowels were opened that day, she replied in the negative, while her nurse had recorded two movements on her chart; again she was unable to name properly the articles of diet which she was taking, always giving the names of different foods which she had never had.

About September 27 she began to have periods of excitement, during which she would shout terrible oaths and use very vile language. On October 6 she said she had been up and around all day, and had been down town in the morning; at the same time she was scarcely able to move in the bed. She complained of much pain in her hands, but otherwise said she was feeling pretty well.

On November 7 while the interne was in the ward taking some notes, the patient said she was not able to go down town to have her pictures taken, but would go tomorrow. She talked in a numbling or muttering manner, which could seldom be understood, and for this reason many of her delusions could not be recorded.

About September 25 she developed incontinence of urine and feces. Tenderness over the extremities had become almost unbearable. There was complete double wrist drop, and she had become somewhat wasted both in the extremities and about the face. On November 11 the above symptoms were all present and exaggerated. On lateral excursions of the eyes nystagmus was produced.

On October 10 Dr. Shumway reported extraocular movements good in every direction; the pupillary reactions prompt and that ophthalmoscopic examination showed a perfectly normal eye ground on each side.

The examination of the organs of the chest and abdomen gave no points that would be of any value in recording the nervous and mental phenomena of the case.

The muscles of the hand, especially the dorsal interossei, were slightly wasted; otherwise the upper limbs appeared well developed. Motion was present in all directions; resistance to passive movements was much decreased in both upper extremities; wrist drop was present on the left side, not on the right. The grasp of both hands was diminished, slightly more so on the left side. The biceps, triceps, and wrist reflexes were absent on each side. Pressure over the course of the nerve trunks of both upper limbs caused pain, and sensation to touch and pain appeared to be about normal in both.

Neither lower extremity showed any atrophy, and motion was greatly lessened in both, the patient being able only to bend the knee to a small angle, and unable to raise the feet from the bed.

The above notes by Dr. Carpenter were finished about five or six weeks before the death of the patient, and after that time nothing of much importance for the purposes of study was added to the clinical history of the case. The patient became weaker and

weaker, both physically and mentally, with, however, occasional short periods of slight improvement. She developed a bed sore on the left hip, the incontinence of urine and feces became more marked, the pain on pressure and manipulation of nerves and muscles persisted, and even when she became apathetic to the point of stupor, was sufficient to arouse her with evidences of distress.

Bedside notes continued to be taken at intervals. A tendency to excitement was noted on one or two occasions. Several records were made of extreme tenderness of the muscles and also of tenderness and pain in the limbs. Small swellings appeared on the arms, the forearms, feet, and other places, these apparently being connected in some way with the inflammation of the nerves. Her temperature rose and fell several times. The day before her death the temperature, pulse, and respiration were all increased. The lungs were full of coarse rales. She died December 31.

A general necropsy was made a few hours after death, practically all the organs of the body being examined. The pathological diagnosis included chronic pulmonary tuberculosis, chronic pleurisy, fatty degeneration of the heart, fatty, cirrhotic, and congested liver, chronic catarrhal cholecystitis with cholesteric gall stones, fibroid pancreas, and chronic diffuse nephritis.

The scalp was found unusually adherent to the calvarium. The skull was hard and of normal thickness; the diploic surface was diminished; the dura was free from the calvarium; the brain weighed 1240 gm. (39.85 ounces); the brain showed normal external configuration with fairly marked edema of the pia and venous congestion. The lateral ventricle showed nothing abnormal. The brain was placed in formalin solution for further study, as was also the cord, retained within its membranes. The left sciatic with its two main subdivisions (internal and external popliteal), the right posterior tibial, the right external plantar along the outer margin of the foot, the right internal plantar to the great and second toes, and a section of the pneumogastric nerve were also preserved.

In this case microscopic sections were made from the lumbar and cervical portions of the spinal cord; and from the oblongata and the pons. In the cerebrum sections were made from the paracentral lobules, the angular gyre, and the mid-precentral, occipital, calcarine, and temporal regions. Sections were also

made from pieces of muscle, and from the external plantar, the sciatic, the internal plantar to the great toe and the pneumogastric nerve. In addition, sections were made of the anterior half of the entire cerebral hemisphere.

These various sections were stained by the Nissl, the Weigert hematoxylin, the alum-hematoxylin, acid-fuchsin, and in some cases the Marchi methods.

Sections from the lumbar region showed considerable chromatolysis of the nerve cells of the anterior horns, with peripheral displacement of the nuclei. This chromatolysis seemed to invade the whole of the cell body. Some of the cells were greatly vacuolated. In three serial sections from the mid-lumbar region, an accumulation of round cells was seen lying immediately in front of and mesal to one of the anterior horns. These cells were both polymorphonuclear and mononuclear in type. In the midst of this accumulation was seen a dense body, staining with Nissl method dark blue-black. Under the oil immersion objective, this was found to consist of an accumulation of cocci. Near the periphery of this dense mass was seen what suggested diplococci. A blood vessel could be seen running from the periphery of the cord toward this cellular infiltration.

The cervical region showed about the same alterations as regards the anterior cornual cells, with the exception that the change was not so pronounced. The white matter of the lumbar and cervical regions showed no degenerations by the alum-hematoxylin and acid-fuchsin method. Neither was there any system degeneration as shown by Weigert's method, the columns of Goll, Gowers' columns, the direct cerebellar tracts, the crossed and direct pyramidal tracts, etc., being particularly examined.

In the paracentral lobules were small accumulations of cells, both polymorphonuclear and mononuclear in type, which were apparently not in relation with blood vessels, although it is possible that blood vessels might be found leading to this accumulation in sections either immediately above or immediately below the area examined. Some very small, deeply staining cocci, like those noted in the lumbar cord, were also found in these infiltrations. Some of the cells of Betz were much degenerated and showed central chromatolysis and peripheral displacement of the nucleus.

In the right temporal region there were also found collections of polymorphonuclear and mononuclear cells as noted above. In

this region there were associated close aggregations of red blood corpuscles together with the same cocci-like bodies.

In the right calcarine region, some of the blood vessels of the pia, cut transversely, showed in addition to the thickly packed erythrocytes, a goodly number of polymorphonuclear cells and cocci-like bodies within the lumen.

The right angular gyre, the right mid-precentral, and the right occipital region appeared normal

In the muscle the connective tissue was greatly increased, both between the muscle bundles and between the individual muscle fibers. The muscle fibers were much atrophied. The small nerve fibers within the muscle were much degenerated. The external plantar nerve was greatly degenerated, and the connective tissue about it was thickened. The sciatic nerve was intensely degenerated, as were also the pneumogastric and the internal plantar to the great toe.

The Weigert hematoxylin preparation of the anterior half of the cerebral hemisphere showed no degeneration of the fronto-thalamic band in the anterior limb of the internal capsule, or of the pyramidal tracts in the posterior limb.

As infection was present in this, as in the next case, it was important to know if foci of suppuration were found at the general necropsy. We have already given a digest of the anatomical findings. In addition to this account, the records show that the omentum was large, covering the intestines, and adherent to the sigmoid flexure and brim of the pelvis. In the latter adhesion was a small mass of fat about the size of a large nut, dark red, showing on incision, a small point of suppuration in the interior.

CASE II.—A. C., thirty-two years old, white, was admitted to the wards of Dr. Mills, at the Philadelphia General Hospital, October 16, 1904, and died November 20, 1904. Her father died two years before her admission of carcinoma of the stomach; two sisters died of puerperal sepsis. Her husband denied all venereal disease.

For more than ten years the patient had been a hard drinker of both beer and whiskey, getting drunk about twice a month. Four years before her admission to the hospital she was delivered of a still-born child. In the next two years she had two living children, each dying under one year of age. Seven weeks before admission she was delivered of a seven months' child, which was also still-born. Prior to the onset of labor she was troubled with vomiting and was drinking her usual amount of alcoholics, which was said to be about eight glasses of beer and a half-pint of whiskey daily.

She was in bed for about two days and was then around for two days, when she was compelled to go back to bed because of swelling and pain in both feet and legs. Loss of power in her hands and arms and swelling of her face soon followed. Her legs were painful below the knees, especially on their inner aspects. Her bowels were regular and her appetite remained fair.

About three weeks after the swelling of the feet was first noticed she began to ask foolish questions and make irrational remarks, such as inquiring whether her father, who had been dead for two years, was still in the house and whether he had gone to work; she also asked that her sister, who was also dead, should be sent upstairs to see her.

Examination showed the patient to be a well-developed woman, with slightly flushed face, apathetic expression, and semi-closed eyelids. She turned her head occasionally from side to side, but was otherwise perfectly quiet, lying with her feet turned out, legs straight, fingers slightly flexed upon the palms and hands upon the forearms. She muttered occasionally about members of her family and made irrelevant remarks. She answered questions with fair intelligence, however, and did as she was told; she lapsed at times into sleep. Her gaze was generally fixed; her pupils were small and equal, but reacted sluggishly, if at all, to light, and but slightly to accommodation. Ocular movements were good in all directions.

The tongue was protruded evenly without difficulty and was not tremulous, but presented a considerable white coating and a stringy mucous.

Her right forearm could be feebly flexed against resistance and her left was even weaker. The grip of the right hand was poor; that of the left was still less. Neither hand could be extended on the forearm. Sensation to pain and touch in the upper extremities appeared to be preserved. Pressure over the bellies of the muscles of the arms caused pain, as did also rapid passive movements. The reflexes were abolished.

The legs could be slightly flexed, and the feet slightly rotated. Otherwise the woman was powerless to move herself in bed. Manipulation of the feet, as in efforts to elicit ankle clonus, caused pain, as did also grasping the calf or thigh muscles. The knee jerks were both absent as were also ankle clonus and the Babinski response.

Expansion was good on both sides of the chest. The apex beat was not visible. The pulse was small, of poor force, and slightly accelerated. The cardiac impulse was feebly felt in the fifth interspace in the mid-clavicular line. An area of dullness extended from the third rib to the left edge of the sternum and to one-half inch outside of the mid-clavicular line. The percussion note was good over both lungs and the breath sounds were clear. The liver dullness began above the fifth rib, but the organ could be felt below the costal border; the spleen could not be felt below this border.

This woman's physical and mental condition did not change much during the time she was in the hospital, except that it got worse during

the last few days of her life. She continued to be helpless in her limbs and to have pain, both subjective and on handling them so as to compress the nerves and muscles.

She was put on the usual treatment for multiple neuritis consisting of the internal use of small doses of mercury and of the salicylates, iodides, and bromides, her other treatment being symptomatic and chiefly remedies to relieve pain and produce sleep.

She died November 20, two days before her death developing great weakness, which passed into stupor.

In this case, as in the preceding case, sections were made from the lumbar and cervical portions of the spinal cord and from the oblongata and pons; also in the cerebrum from the paracentral lobules, the angular gyre, the midprecentral, occipital, calcarine and temporal regions. In addition sections were made from muscle tissue and from the trigeminal, abducens, and facial nerves; also, as in Case I, from the anterior half of the entire cerebral hemisphere. The sections from this case were stained in the same manner as those from Case I.

The sections made from the lumbar region showed great degeneration of many of the anterior cornual cells, this being evidenced by a disappearance of dendritic processes, by peripheral displacement of the nucleus, and by central chromatolysis. The changes in the cervical cord were the same as in the lumbar region, although possibly less intense. In the gray matter of the anterior horn were seen several small hemorrhages. In this case, as in the previous one, the examination of the Weigert preparation of the anterior half of the cerebral hemisphere proved negative; the fronto-thalamic band and pyramidal tracts were normal.

By the Marchi method the oblongata and the lumbar cord showed no typical degeneration. Sections of the oblongata stained by the Nissl method showed a number of hemorrhages in the posterior part. In this locality was also found a very pronounced perivascular round cell infiltration. There was no round cell infiltration of the pia; it was remarkable that this perivascular round cell infiltration within the oblongata should be so sharply confined to one or two vessels and should be so intense. At a higher level, was seen a slight round cell infiltration in the pia.

In the anterior part of the pons was a collection of round cells in close juxtaposition to a blood vessel. This collection was similar to those found in the previous case.

The paracentral lobules in this case seemed normal, as regards

the nerve cells, but the pia was considerably thickened, and along its outer edge were found occasionally proliferation of endothelial cells forming distinct masses like those described by Spiller* in a case of lead encephalopathy. The pia was in many places firmly adherent to the cortex and presented an increase in the number of nuclei. The calcarine, precentral, angular gyre, the occipital and frontal regions presented no abnormalities.

Sections of the trigeminal nerve stained by the Marchi, the acid-fuchsin, and the Weigert method appeared normal.

As the only post-mortem investigation made in this case was of the nervous system and some of the muscles, we have no facts regarding the existence of purulent foci in any part of the body.

Case I was clearly one of advanced polyneuritis. The nerves of both the lower and upper extremities were extensively involved in the inflammatory and degenerative process, as shown by the symptoms during life and also by the microscopical investigation. Some of the muscle masses were also tender to pressure and the pathological examination showed degeneration of the muscle fibers as well as of the nerves entering the muscles. While it was not possible, owing to the condition of mental excitement and degradation present, to develop many of the typical symptoms of the polyneuritic psychosis, sufficient was obtainable to make justifiable the diagnosis of Korsakoff's disease. For a time the patient was in an excited, almost maniacal, state. A few pseudo-reminiscences or memorial fabrications were noted, and her mental condition was evidently one of extreme delusion and confusion, obscured somewhat by her low state of vitality.

The disease had developed during pregnancy and came to its height shortly after delivery, illustrating the fact to which attention has already been called that the disease not infrequently occurs as the result of some special infection or poison in alcoholic patients, the chronic alcoholism having produced the condition of non-resistance.

The toxic effects of alcohol are, however, sufficient in themselves to establish the disease. In a long experience with cases of multiple neuritis, not a few of which have shown mental symptoms of the Korsakoff type, we have seen a considerable number

* Spiller, Wm. G., *Journal of Medical Research*, 1904.

of cases in which alcohol was the only ascertainable etiological factor.

In Case II the multiple neuritis developed in connection with the puerperal state, the patient having been delivered of a still-born child seven weeks before admission. The swelling and pain in her feet which inaugurated the disease came on a few days after the premature delivery. The physical symptoms present were clearly those of multiple neuritis, such as pain, tenderness to pressure over nerves and on handling muscle masses, paralysis and lost deep and superficial reflexes. Owing to the rapidity with which the case passed into an apathetic and then stuporous state, the mental symptoms noted were not numerous. She showed, however, impaired memory for recent and remote events, a confused and delusional state, and disorientation. She talked about her father and her mother, who had been dead for some time, as if they were present in the house.

The findings in this case, so far as the nerves, muscles, and spinal cord were concerned, were similar to those in Case I. The cells of the cerebral cortex which were studied in various regions as indicated in the report did not present the same post-inflammatory and degenerative conditions which were noticeable in similar areas in Case I. The pia, however, showed more involvement and was more adherent.

In both cases here considered the muscle fibers were degenerated as in the case described by Cole, who holds that it is probable that the changes in muscle fiber are due to the direct action of toxin on true muscular tissue; that the interstitial fibrosis often present is an end product and not a primary affection. He also believes that one reason why the nerves in the limbs are more affected in the disease is because of their passage through muscle masses in which they are more bathed in poisonous lymph. Our experience, not only with the cases under observation, but with many others observed at the Philadelphia General Hospital and elsewhere has been that grasping muscle masses or otherwise manipulating them usually causes extreme pain; as much indeed as that caused by more direct pressure upon more exposed nerves, as for instance, upon the nerve trunks in the popliteal space or in the foot.

A study of our cases would seem to show that the neuron as a whole was attacked by poison causing the disease; not its distal or

medial portion or the nerve cells alone. The conditions present in the peripheral distribution of the nerves and in the cell bodies in the ventral horns seem to clearly point to this conclusion. In a paper on Landry's paralysis, written in collaboration with Dr. Spiller by one of us (Dr. Mills),[†] this same conclusion was reached, namely, that in some cases at least, the neuron as a whole was attacked by the toxin of the disease, and not simply its periphery or its central portion.

The Argyll-Robertson pupils were not present in our cases—a fact of not as much diagnostic importance as in cases of the so-called neuritic pseudo-tabes—cases able to go around, but showing decided paresis and ataxia. The absence of the symptom, however, in cases of such severity indicates its diagnostic value in other more doubtful cases. Cole properly makes a strong point of the absence of the Argyll-Robertson pupil in pseudo-tabes, believing that more than any other symptom it separates this disease from true tabes.

We shall content ourselves with the presentation of clinical histories and pathological findings in these cases and a brief reference to a few of the more recent contributions on the subject of the pathology of the polyneuritic psychosis. The results obtained in the study of our cases can be compared with advantage to those detailed in these contributions. The microscopical findings in our cases would seem to bear out some of the most important conclusions of Cole. This writer made a careful study of three cases of chronic alcoholism with neuritic disorders and changes in the central nervous system.

These observations, according to him, suggested the following: (1) That the peripheral and central lesions express a nervous degeneration of toxic origin, in the production of which no essential part is played by changes in interstitial tissues supporting the nervous structures, or by changes in the blood vessels concerned with their nutrition. (2) The changes in the nerve-cells are not the mere results of antecedent damage of nerve fibers, but the changes in the fibers and cells together express a highly selective affection of whole neurons. (3) The peripheral neuritis is simply a local expression of this affection, and is not of purely local and peripheral causation. (4) The lesion of peripheral

[†] Mills, C. K. and Spiller, Wm. G., *Journ. of Nerv. and Ment. Dis.*, June, 1898.

neurons is only one of many manifestations of the disease, and is accompanied by lesions identical in nature, affecting many groups of neurons situated entirely within the central nervous system. (5) The central changes are not attributable to the peripheral neuritis, and though in some cases the peripheral neurons are mainly affected, in others the morbid process chiefly implicates central neurons; but these two groups of cases do not appear to be sharply divided.

With regard to the mental disorder in Korsakoff's disease, Cole says that this is probably related in some way to the cortical changes, an opinion with which we accord.

In Cole's study of his three cases is given an interesting table in which is shown at a glance the changes in the peripheral nervous system, the central nervous system, and the vessels.

Sims has recorded the anatomical findings in two cases of Korsakoff's disease. In the first of these two cases the most important findings were:

Slight arteriosclerosis, hypostatic pneumonia, fatty infiltration of the liver, acute degenerations of many of the peripheral nerves, axonal reaction in cells of the anterior horns, Clark's columns, and many cranial nerve nuclei, degeneration in the posterior columns, direct cerebellar tracts, and the root bundles, and moderate acute alteration of the cortical cells.

In the second case they were:

General arteriosclerosis involving the aorta and coronaries, fatty degeneration of the heart, liver, and kidneys, acute bronchitis, acute degenerations in the peripheral nerves of the lower extremities, and also in the vagi, axonal reaction in cells of the anterior cornua, in Clark's columns, some cranial nuclei, and the Betz cells of the cortex; also, acute degeneration of the cortical radiations, and of both motor and sensory systems of the cord, as well as degeneration of the cord not easily reconcilable with the systemic changes. Vascular changes in the cord and cortex, with numerous miscellaneous hemorrhages throughout the cerebrum were also found.

In the *Archiv für Psychiatrie und Nervenkrankheiten*, Dr. Wehrung has reported thirty-four cases of the Korsakoff symptom-complex. The following is a digest of his findings:

In the spinal cord the following changes were noted: Ganglion cells rarer, shriveled and processes lost, slight anterior poliomye-

litis, congenital changes in the central canal, proliferation of connective tissue in the column of Goll, proliferation of connective tissue in right lateral column, ectasia and hyperemia of vessels of the cord, vacuoles in ganglion cells of anterior horns, increase in nuclei of lining of central canal, partial degeneration of column of Goll, calcification in membrane of spinal cord, hemorrhages in white substance of cord, anterior cornual cells stain more intensely than normal, disseminated degeneration of nerve fibers, central chromatolysis and excentric nucleus, hydromyelia, degeneration of root zone of posterior column, pigmentation of cells of anterior horns, proliferation of neuroglia around anterior horns, and slight perivascular proliferation of nuclei.

In the oblongata, pons and brain generally, the following changes were observed:

Granular atrophy of pyramidal cells of the cortex, pia oedematous, chronic arachnitis, decrease of tangential fiber net work especially of frontal convolutions, area of softening in gray matter of cortex in motor zones, area of softening in left island of Reil, extensive hemorrhagic encephalitis in the gray matter in the third ventricle, atrophy of corpora mammilaria, hemorrhagic changes in distal nucleus of vagus, calcification of vessels in brain, hemorrhagic encephalitis in thalamus, softening in corona radiata, granular condition of ventricular ependyma, hemorrhages in lateral ventricle, hemorrhages in centrum ovale, hemorrhages in nucleus caudatus, myxosarcoma of right temporal convolution, sarcoma of third ventricle with metastasis to fourth, all stages of central chromatolysis in paracentral lobe, tumor of the base, syphilitic encephalitis, and severe gliosis.

In order not to mislead, we would especially call attention to the fact that no one case in Dr. Wehrung's^{*} paper embodied more than a few of the lesions noted in these tables.

Reviewing the findings of Dr. Wehrung, it is evident that while many of them fairly represent the pathological anatomy of polyneuritis and the polyneuritic psychosis, many others are to be regarded as simply coincident or accidental lesions, having no relation whatever with the disease under discussion.

^{*}Wehrung, Gaston, Beitrag zur Lehre von der Korsakoff'schen Psychose mit besonderer Berücksichtigung der pathologischen Anatomie. Ein Weiterer Fall. Archiv für Psychiatrie und Nervenkrankheiten, Vol. XXXIX, 1904-5, p. 679.

THE DISTRIBUTION OF THE MOTOR ROOT IN THE ANTERIOR HORN.

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IN a short but comprehensive paper by Blumenau and Nielsen¹ the relation of the posterolateral cell groups of the anterior horn of the fifth, sixth, seventh, and eighth cervical and first thoracic segments of the spinal cord to the muscles of the shoulder is exploited. Mention is made of experimental work on animals along these lines by Marinesco,² M. and Mme. Parhon,³ and De Neef⁴ and a similarity of cell-group functions between man and the lower animals is shown.

The case I am reporting has already been made the subject of a study in another line by Spiller,⁵ to whom I am indebted for the pathological material.

A patient denying specific disease and having suffered from severe left frontal headache for two years began to show signs of brain tumor, viz., motor aphasia, weakness of right arm and hand, partial paralysis of right side of face, and convulsive seizures in which consciousness was not lost. "A diagnosis of tumor of the lower left motor area was made. It was supposed to affect Broca's area by pressure or to invade it slightly, inasmuch as the motor aphasia was

incomplete." At the operation (performed by Dr. C. H. Frazier) the tumor was exposed, and because of the extent and vascularity no attempt was made to remove it in its



FIG. 1.—Spinal cord showing small tumor involving the greater part of the right anterior nerve root of the sixth cervical segment.

entirety. The tumor grew rapidly after the operation, and the patient died about twelve weeks later. At the necropsy "a tumor was found in the posterior part of the left second frontal convolution extending into the middle of the pre-central convolution and to the upper part of Broca's area. The growth was infiltrating and at no part sharply defined from the surrounding tissue. It was, as already said, a small spindle-celled sarcoma."

Upon opening the spinal dura there was found a small tumor involving the right sixth cervical anterior nerve root. This is well demonstrated in the picture (Fig. 1), where is also seen a fasciculus at the lower border of the segment escaping the neoplasm and entering the spinal cord without interference. This growth was entirely unsuspected owing to the masking of any possible symptoms by the partial hemiplegia. Microscopic study shows this small tumor to be a fibrosarcoma.

Serial sections were made throughout the upper part of the sixth segment and one in every five was stained by von Lenhossék's method (thioninphenique).

In describing the changes found in the anterior cornual cells I shall follow the grouping as accepted in the work of F. Sano.⁶

In referring the cell groups of this study to the classification adopted by Sano, I do so because his diagram (p. 32 of the original article) so perfectly illustrates the groups of cells degenerated; but the verity of his conclusions I am in no position to affirm or deny.

The number of cells in each anterior horn was counted, great care being taken not to know which horn was under consideration until after the count for that section was completed. This precaution was taken in order to obliterate as much as possible "the personal equation." The sections are numbered from above downward and the figures are arranged in the following table:

No. of slide.	Right.	Left.	Difference.
5	34	54	20
10	16	29	13
15	35	44	9
20	21	37	16
25	19	45	26
30	29	33	4
35	32	35	3
40	37	49	12
45	25	40	15
50	33	50	17
55	34	51	17
60	41	53	12
65	31	48	17
70	34	55	21
75	38	41	3
80	40	47	7
85	23	33	10
90	52	45	*7
95	32	35	3
100	37	39	2
105	30	36	6
110	38	42	4
115	44	38	*6
120	46	35	*11
125	33	40	7
	834	1054	220

In every section, with the exception of Nos. 90, 115, and 120, there are fewer cells in the right anterior horn than in the left. The greatest differences are encountered in the upper part of the segment. The three exceptions above noted are in the lower part of the tissue examined where the horns are more equal in count, and asterisks are placed in the fourth column indicating these exceptions.

It must be remembered that a small fasciculus of fibers, the lowest in the root, escaped involvement in the tumor mass, and this accounts for the discrepancy in count being less the lower down the section.

Directly opposite the entrance of the nerve root the anterior cornual cells show a marked degeneration in that group which Sano designates as the group for the serratus magnus.

This collection of cells is, in this segment, the most anterior group, and is numbered in Sano's diagram "4." (This diagram has been reproduced in M. Allen Starr's *Organic Nervous Diseases*.)

The sections, as we pass upward, show a change in the locality of the degenerated cells, the anterior Group "4" clearing up and becoming normal as the two groups on either side of and very slightly posterior to it present positive, though not very pronounced, signs of degeneration. Sano indicates these two groups as follows: That on the outside of Group "4" he numbers Group "6," and designates its sphere of activity as the shoulder muscles; that on the inside he calls Group "1" and ascribes to it the innervation of the extensors and rotators of the vertebral column.

The clearing up of the degeneration in Group "4" as we proceed downward is not followed by such pronounced signs of involvement of the two groups, on either side of and posterior to it, as is the case in the upper part of the segment. But, nevertheless, there is the same tendency for the involvement of the group to the outer side, called Group "7," to which is ascribed the innervation of the flexors and pronators, and the group on the inner side which is a continuation of the original Group "1."

By way of a conclusion I offer the accompanying diagram as a possible distribution of the fibers of the sixth cervical anterior nerve root (Fig. 2).

According to this scheme the ultimate destination of the axones of cells in Group "4" below and above the degenerated area is unaccounted for, as is also the middle areas of the mesial and lateral columns.

In a case of brain tumor, where the character of the neoplasm is such that metastasis may occur, it is well to realize how completely spinal symptoms, due to a metastatic growth, may be masked by the hemiplegia present. Should a tumor involve only the anterior root, it is entirely conceivable that no symptoms would be detected even though the patient

had no masking hemiplegia. If, for instance, the sixth cervical anterior nerve root on the right were cut or destroyed by any lesion, there is no muscle which would not still have representation in segments above or below this, with the possible exception of the pronator radii teres and the flexor carpi radialis.

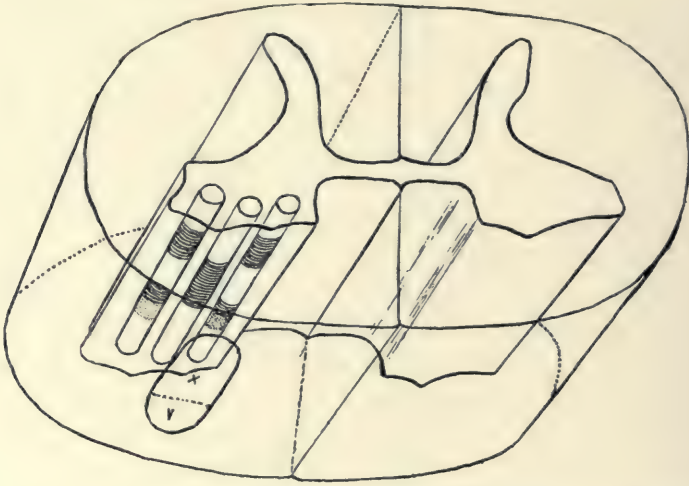


FIG. 2.—A scheme representing a possible distribution of the fibers of the anterior nerve root in the anterior horn of gray matter (the shaded and dotted area). *X*, in the oval, represents the part of the root invaded by the tumor; *Y* represents that part of the root free from tumor. The shaded portions of the three columns represent the locality of the degenerated nerve cells.

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BRAIN TUMOR WITH JACKSONIAN SPASM
AND UNILATERAL PARALYSIS OF THE
VOCAL CORD, AND LATE HEMIPAR-
ESIS AND ASTEREOGNOSIS.

THE GROWTH SUCCESSFULLY LOCALIZED AND REMOVED.*

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As the following case presents one or two points in symptomatology and diagnosis of much interest, as the operation for the removal of the cerebral tumor was entirely successful, and as the neoplasm was of unusual type, we believe that the history of the case is worthy of presentation to this section and also of being permanently recorded.

The patient was a physician in active practice and a man of marked ability in his profession. The case was seen by Dr. Atlee and later was referred to Dr. Mills, who had charge of the patient for several weeks at the University Hospital, at which time the diagnosis of a cerebral tumor was considered highly probable, although it was believed that the recurrent laryngeal paralysis might be due to a peripheral lesion. The case was seen in consultation by several well-known physicians and was thoroughly studied in all its aspects. An account of the various examinations made will be included in the general history which follows:

The patient, a physician, 43 years old, married, had an excellent family and personal history. He had had no illness since childhood except one attack of influenza six years before the onset of his cerebral disorder. He had, however, from childhood, some difficulty in his gastric digestion,

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amounting to slight distress after eating. He had never in his life vomited. He was somewhat troubled with constipation, and had had some catarrhal affection of his throat. For several years he had had frontal and occipital headache.

In March, 1905, a hoarseness was observed for the first time, this persisting for about two months, until the warm weather began. In September, 1905, he had numbness of the left side of the lips, face and of the neck and throat. No twitching of the muscles of these parts, however, at this time was visible, although the patient said that he felt as if the left corner of his mouth twitched.

In January, 1906, the hoarseness returned and the sensation of numbness and twitching reappeared.

In June, 1906, while in a store, sitting, he was taken with a cramp or spasm in the left foot and leg. In a few minutes the spasm extended up the leg and body until it involved the arm, with resulting contractures. He then lost consciousness and fell, remaining unconscious about twenty minutes. When he regained his consciousness he was simply a little confused; he did not vomit, had no nausea, and did not pass into a sleep or stupor. Medical attention to bowels and stomach aided him for some weeks. His minor facial attacks previously mentioned also got fewer and less severe.

About eight or nine weeks after the first severe convulsion with unconsciousness, he had another similar attack. He felt the cramp or spasm in the leg and had time to lie down before he became unconscious. The spasm took the course of the preceding one, and he remained unconscious for ten or more minutes. He never had any tingling, formication, or aura of any kind with these convulsions. His facial attacks were mere spells of numbness with inability to speak, so that he had to motion for what he wanted. Others around him did not observe any strabismus or any twitchings or contractions of the face in these spells. He was placed on bromid and iodid, and on Sept. 10, 1906, he was referred to Dr. Mills, under whose care he remained from this date until Oct. 16, 1906.

He was carefully examined for symptoms referable to nervous disease on various occasions while in the hospital of the University of Pennsylvania. No extra-ocular palsies were present, and no paralysis could be determined in the distribution of either the fifth or the seventh nerves on either side, although the naso-labial fold seemed to be deeper on the left than on the right. Paralysis or paresis was entirely absent in the limbs on each side. The deep reflexes, both in the upper and lower extremities on both sides, were prompt, but more so on the left. This was especially so of the triceps jerk and of the knee jerk. Ankle clonus was absent on each side. Plantar stimulation on the right side produced plantar flexion; on the left the toes remained stationary. Sensation for touch, pain, and temperature was normal all over the

body, as was also the sense of location. With the hand absolutely relaxed, the patient did not seem to recognize when the left thumb and forefinger were moved. When the left thumb and forefinger were moved he could not place the right thumb and forefinger in the corresponding positions. Astereognosis was not present, as determined by examination with a number of different objects.

Dr. G. E. de Schweinitz reported that his ophthalmoscopic examination showed no optic neuritis. Dr. E. A. Shumway reported that the patient had normal vision in each eye; that the pupils were equal, movements prompt, convergence poor, the right eye diverging, other movements being good; that both the nerves, especially the left, showed slightly blurred upper and inner margins, probably not indicating a true neuritis; visual fields normal and moderate arteriosclerosis of retinal vessels.

Dr. George R. Rohrer, of Lancaster, sent with the patient a report of his ophthalmoscopic examination, which he said revealed an indistinctness of the optic nerves, upper, nasal and lower portion, in both eyes, and a slight contraction of fields, which when taken in connection with his attacks would indicate central disturbance.

Owing to the paralysis of the recurrent laryngeal nerve, Dr. John H. Musser was asked to make an examination of the chest, which was supplemented by an x-ray investigation. He reported that he found enlarged post-cervical glands, a difference in the pulses, a high-pitched percussion note to the left of the sternum, a murmur along the aorta, and a rapid pulse. It seemed to him that there was mediastinal pressure, probably due to enlarged glands, although possibly to an aneurism. The Roentgen ray examination showed a shadow about the bronchi, as of enlarged glands.

Dr. B. A. Randall made a laryngeal and also a nasopharyngeal examination of the patient, reporting as follows: The hoarseness is from paralysis of the left vocal cord; the voice is stuffy; fauces and pharynx vault are much thickened and red; the left cord and arytenoid immovable near the median line. The examination at which the above notes were taken was made on Sept. 20, 1906. Under the same date Dr. Randall also reported: "The patient shows no measurable loss of hearing in either ear, although he gives distinct preference to the left as compared with the right. His bone conduction seems normal and fairly to exclude nerve lesion on either side. My glance at his nose and throat is also negative, with no tubercular or other evidences to account for the paralysis." May 10, some months after the operation, Dr. Randall's examination showed that the cord was atrophic-looking near the median line.

Although a tumor involving the right facial area was considered probable, as the general symptoms of brain tumor were absent, and as there was some probability that the paralysis

of the inferior or recurrent laryngeal nerve was due to mediastinal disease, as glandular enlargement or aneurism, it was concluded for the present to postpone an intracranial operation, the understanding being that the question of operation would be taken up later should additional symptoms of focal cerebral lesion appear. The patient returned to his home near Lancaster, where he was again seen by Dr. Atlee.

After his return headaches appeared in the occipital region, usually coming on at night, awakening him at times and requiring morphin hypodermically for relief. They were worse when lying down, were relieved by an upright posture, lasted several hours, and appeared at about weekly intervals.

Weakness in the left side appeared about Dec. 1, 1906, and failure of word memory both in speech and writing, was noticed a few weeks later. The hoarseness and sensation of twitching continued 1 this time.

When examined, Jan. 26, 1907, he had marked weakness of the entire left side, including the tongue, which was protruded to the left. The deep reflexes were accentuated on the left side. Sensation to pain, touch, and temperature was very little, if at all, dulled. There was astereognosis on the left side and failure of word memory. The hoarseness amounted to entire loss of voice.

On Feb. 1, 1907, following strenuous attempts to continue in practice and realizing his inability to do so, the patient was in bad shape, pulse 120 and weak, and much depressed mentally. He could hardly remember any words at all; he had no power of phonation; the entire left side was almost powerless; astereognosis was complete in the left hand; and sensation was impaired. He was ordered to bed at absolute rest and one week later entered St. Joseph's Hospital, Lancaster, Pa., much improved.

On Feb. 9, 1907, examination showed the temperature subnormal, varying from 97 to 98; the pulse rather rapid, averaging 100. The heart, lungs, abdomen and kidneys were examined without result. The patient had a cough, not accompanied by any expectoration and apparently of laryngeal origin.

The entire left side of the body had diminished power, and the tongue was protruded to the left. Except for weakness, the gait and station seemed good. The deep reflexes were accentuated on the left side, normal on the right. Sensation was impaired very slightly on the left side, if at all, but astereognosis was marked, although not complete, in the left hand. Coordination and localization seemed normal on both sides.

The left vocal cord showed adductor paralysis; both the cord and the entire larynx were much congested, apparently due to the cough. The vessels in the left retina were dilated, but no neuritis and no disturbance of vision were present. The vessels of the right retina were not dilated.

At this time the patient stated that he had a sensation of pulsation in the parietal region, synchronous with the heart beat. There was no tenderness anywhere on percussing the cranium.

The diagnosis of a tumor of the cerebrum, originating at the lower end of the precentral convolution and involving a large part of this convolution, and to some extent the parietal lobe as shown by the presence of astereognosis, being now considered practically certain, in spite of the absence of the general symptoms with the exception of recent headache, it was decided to operate.

The operation was performed by Dr. Atlee at St. Joseph's Hospital, Lancaster, Pa., Feb. 12, 1907. Ether was chosen because the anesthetist was especially skillful in its use. Its administration was preceded one-half hour by morphin, gr. $\frac{1}{2}$, and atropin, gr. $\frac{1}{100}$, given hypodermically.

The fissure of Rolando was located both by Chiene's method and from Reid's lines, and an osteoplastic flap planned to expose the region around the lower end of this fissure and the anterior part of the parietal lobe was made. The thickness of the skull, however, one-half inch, was too great for the bite of the DeVilbiss rongeur, which Dr. Atlee had always before found to be a very satisfactory instrument, and this plan had to be abandoned. Therefore the bone was simply removed with rongeurs until a triangular area was exposed extending upward along the line of the fissure for $1\frac{1}{2}$ inches, and back an equal distance with the base parallel to Reid's base line. The dura was tense, bulging, and did not pulsate. On incising this membrane a mass at once protruded to the level of the outer table. This mass was not encapsulated and apparently infiltrated into healthy brain, but nevertheless a line of cleavage revealed itself to the finger tip and the tumor was comparatively easily shelled out. Very little hemorrhage followed its enucleation and this was controlled by the gentle insertion of a sponge wrung out of hot salt solution. The dura was closed with catgut, a celluloid plate fitted and the scalp closed with silk worm gut, without drainage.

The operation lasted one hour and no shock followed, the patient leaving the table with a temperature of 98.4 and a pulse of 140, but good.

The mass removed measured $2\frac{1}{2}$ by 2 by $1\frac{3}{4}$ inches. It was oval in shape, not adherent to the dura, but no pia was noticed over it. It was soft in consistency, although distinctly more firm than the brain. It was grayish brown in color, with yellowish areas of fat. The center of the mass was filled with a syrupy liquid containing much brownish detritus and some free oil globules.

The celluloid plate was sterilized by boiling in plain water, and molded while in this boiling water at the table side to accurately fit the curve of the cranium. This procedure is

comfortably carried out when gloves are worn. The plate was not fastened, being simply inserted under the scalp.

Under date of March 9, 1907, the following notes were made by Dr. Atlee: "The patient's convalescence has been aseptie and afebrile. The highest temperature was 101, the highest pulse 140. The temperature reached normal on the fourth day, but the pulse is still a little frequent, about 100. The patient came out of ether bright and smiling and did not vomit. There was little pain, no headache and no convulsive seizures. He sat up on the tenth day, was out of bed and took several steps on the fourteenth day. There were no symptoms of any damage to the brain in removing the mass except some increased weakness of the deep flexors in the left forearm and the adductor pollicis. Otherwise the left side is somewhat stronger than before the operation. His memory for words and stereognosis is good. There has been no twitching sensation in the face since the operation. Phonation has returned momentarily once or twice. About three weeks after the operation mild taradism was ordered to the left side, but in spite of warning it was used too vigorously and resulted in a spasmodic seizure in the left foot, followed in three hours by a second seizure and in twelve by a third. No spasms have occurred since, nor has electricity been used since."

A considerable accumulation of fluid occurred at the site of the operation. Whether this was cerebrospinal which had leaked through the dural stitches or whether it was due to the irritation of the plate could not be said, but in Dr. Atlee's experience this has occurred in all cases in which the plate was used, and has always disappeared in three weeks, as it did also in this case.

The pathologic examination of the tumor removed in this case proved of unusual interest. This examination was made by Dr. William G. Spiller, to whom we are indebted for the following report with comments:

The tumor is friable and contains in its interior much mucoid substance. This disappears when the tissue is placed in alcohol and ether. A small portion of this mucoid material under a cover glass is found to contain round or nearly round bodies, all about the same size, resembling red blood corpuscles, which they probably are. They have probably escaped from the vessels of the tumor. The mucoid material contains also numerous cholesterin plates.

The tumor microscopically presents much the appearance of the chorioid plexus, and contains many convoluted single rows of cuboidal or cylindrical cells like those of the chorioid plexus. When a vessel is cut transversely the cells form one, two or even three rows surrounding the vessel, as seen in a perithelioma. The papillomatous processes are separated from

one another in certain parts of the tumor by mucoid material containing scattered cells, like those forming the tortuous rows and also some red blood cells. The cuboidal or cylindrical cells forming the rows have each a large nucleus staining fairly well, and placed at the inner end of the cell. Usually the blood vessels within these rows of cells can not be detected, but occasionally can be made out. Fibrous tissue is not found between the papillomatous processes. Fatty degeneration is not seen in the cells stained by osmic acid, but

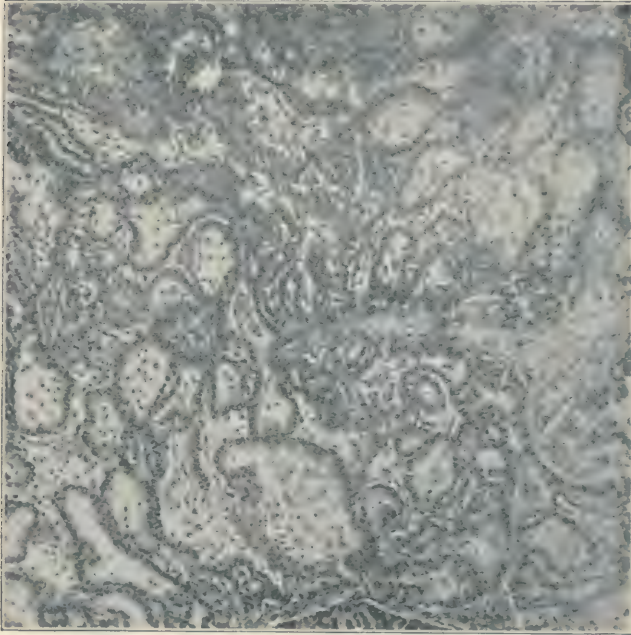


Fig. 1.—Photograph of a microscopic section of the tumor, showing its resemblance to the structure of the chorioid plexus.

in frozen osmic acid preparations, the free ends of the cells have a swollen appearance, as though they were distended with fluid.

It seems probable that the tumor, epithelial in character, grew from the chorioid plexus.

The microscopic appearances presented by the tumor are shown in Figures 1 and 2.

Borst,¹ however, says that perithelioma of the pia may have a peculiar form when it assumes a papillomatous

1. Die Lehre von den Geschwulsten, 1, 347.

character. He mentions that Rindfleisch observed a papilloma of this kind on the cerebellum, between the pia and the left cerebellar lobe. By teasing, the entire tumor separated with a mass of papillæ each containing a central blood vessel, with a double row of epithelial cells about it and very little connective tissue. Borst says he himself has seen such a papilloma arising from the chorioid plexus.

These tumors have recently been discussed by Bielschowsky and Unger,² who say that epithelial tumors of the brain are rare, and when they occur they commonly develop from the chorioid plexus. The findings are usually the same in all these tumors, i. e., a simple hyperplasia of the chorioid plexus. The tumor consists of tortuous vessels with thin walls, having on the outer side of a delicate adventitia one or more rows of cylindrical cells. Where the vessels are cut transversely the epithelial cells are seen radiating from the lumen of the vessel. Interstitial connective tissue is scarce and may be absent. Retrogressive metamorphosis and hemorrhages in the tumor are common. Different names have been given to these tumors; they have been described as hyperplasias (Brüchanow); as papillomata (Saxer, Selke); as peritheliomata on account of the perithelioma-like arrangement of the epithelium about the vessels, and in the attempt to connect them with other tumors arising from the vessels (Lubarsch, Borst). They have been described also as adenomata, inasmuch as the chorioid plexus is a secreting organ. They are relatively benign; they have no tendency to metastasis, or to infiltrate, and are dangerous simply by their size. The loss of brain substance even from the largest of these tumors is relatively slight. Exceptions, however, may occur.

In a case reported by Bielschowsky and Unger the tumor had its origin from the chorioid plexus and extended to the cerebral cortex. Another growth was found in the right cerebello-pontile angle and was connected with the chorioid plexus of the cerebellum. The center of the tumor contained mucoid substance. Microscopic examination revealed numerous small tumors on the cortex in relation with the pia. The mucoid substance found in two large tumors was believed to be a secretion of the cells. In the small tumors the vessels

2. Archiv. für klin. Chirurgie, 1906, lxxxI, 61.

were not solitary, but formed vascular papillæ covered with epithelial cells. The drawing given of these structures by Bielschowsky and Unger resembles closely the findings in the present case. The capillary vessels in some places were united by delicate fibrous connective tissue. In osmic acid preparations amorphous masses and fatty granular cells were found, the latter surrounding blood vessels. Microscopic examination showed that the tumor grew by infiltration, a vessel penetrated the

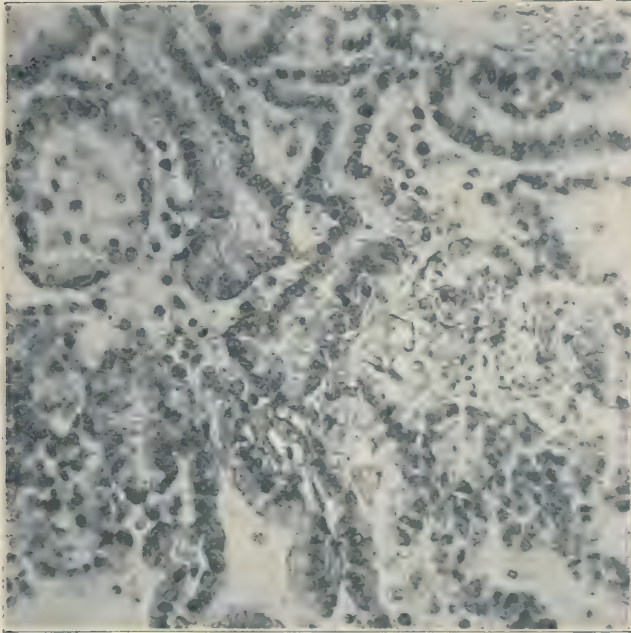


Fig. 2.—The same as Figure 1, only under higher magnification.

adjoining brain substance and the nervous tissue enclosed by it degenerated. The primary process was sprouting out of a delicate capillary and cell lining followed.

In no other case of epithelial tumor of the chorioid plexus reported was such malignancy observed, as shown in the case of Bielschowsky and Unger, by the multiplicity of tumors and their infiltrative growth.

Bielschowsky and Unger mention that peritheliomata of the brain resemble these tumors of the chorioid plexus

closely, and that in some cases distinctions are impossible, and that Lubarsch and Borst do not attempt to separate the two forms; but in this they ignore the fact that the perithelium of the vessels is of mesodermal origin, while the epithelium of the choroid plexus is of ectodermal origin. Bielschowsky and Unger believe that in their case the large cerebral tumor was primary and that some particles were carried from this through the lateral recess of the fourth ventricle and formed the origin of a new growth in the cerebello-pontile angle, from this in turn metastasis occurred throughout the pia by means of the cerebrospinal fluid.

As indicated in the last report made by Dr. Atlee regarding the condition of the patient after the operation, with one exception improvement had steadily progressed. This exception was the occurrence of the spasms, apparently attributable to the use of a strong faradic current. The patient was soon able to go about almost as one who had never been affected by intracranial disease.

On May, 23, 1907, he wrote to Dr. Mills, giving some account of his condition since the operation and at the time of writing. In this letter he said:

I have been out of the hospital about two months; that short time has made a decided improvement. My memory for words and spelling came back at once after the operation. My voice is gradually improving. I could not speak above a whisper while in the hospital, but now I can speak loud enough for anyone to understand. . . . I can eat and sleep well and have gained about twenty-five pounds.³

With regard to several points in focal diagnosis this case, although interesting, proves nothing which has not already been demonstrated by physiologic experiments and clinicopathologic observation. The Jacksonian spasm with its initiating sensory aura, the attacks consisting of a sensory discharge confined to the face, tongue, throat and neck, the late development of astereognosis as the tumor grew, are clinical phenomena all easily explicable on the views which are now receiving general acceptance, that the cortical area for the face

3. The patient kindly presented himself for examination at the meeting at which an account of his case was given, being then in unusually good condition, as indicated in his own report just cited. His voice was considerably improved, but there was still evidence of the unilateral paralysis of his vocal cord. He had good use of the side formerly partially paralyzed, and otherwise was about in the condition detailed in the body of the paper.

and limbs is situated in the precentral convolution and that for stereognostic conception in the parietal lobe; in other words, the parts which were directly affected by the presence of the tumor.

Whether or not the unilateral paralysis of the vocal cord was due to the cerebral lesion is a question more interesting and much more difficult. This paralysis, as determined by the study of the patient's condition and also by laryngeal examinations, had all the features of a unilateral peripheral recurrent laryngeal affection, and, indeed, it can not be positively stated that it was not due to such peripheral disease. It is interesting, however, to recall that the patient's voice, which had disappeared entirely, recovered considerably after the removal of the cerebral tumor, although at the time the last laryngeal examination was made no great change had taken place in the paralyzed and somewhat atrophied vocal cord. It is generally believed that the cortical laryngeal centers have bilateral control, in other words, that the center in each hemisphere controls the movements of adduction and abduction of the vocal cords on both sides. It might be well here in concluding this paper to summarize the most important facts regarding cortical laryngeal centers. It will be seen that a portion of the tumor implicated the exact region in which the cortical center for the larynx is situated, and curiously the tumor was a right-sided one, the centers for the larynx, as will be seen presently, being presumably more developed in the right than in the left cerebral hemisphere.

In 1888 one of us, Dr. Mills,⁴ summarized the most important facts recorded up to that time regarding the cortical centers for laryngeal movements. In 1877 Seguin⁵ reported a case with necropsy with left hemiparesis and impairment of speech and phonation. The patient after the attack could never properly regulate the pitch of the voice. The surface of the right third frontal convolution was degenerated, the same change, but less marked, being found in the corresponding location on the left side. From this observation it has been sup-

4. Mills (Chas. K.): *Cerebral Localization in Its Practical Relations*, Trans. of the Congress of American Physicians and Surgeons, I, 1888; also *Brain*, xii, July, 1889.

5. Seguin (E. C.): Trans. of the American Neurological Association, 1877, p. 102.

posed that the center for laryngeal movements might be located in the right third frontal.

Krause⁶ in 1883 found that on excitation of the cortex of the dog in a region corresponding to the anterior portion of the lower extremity of the motor region in man rise of the larynx, and movement of the local bands, to a position midway between expiration and phonation, lifting of the palate, contraction of the constrictor pharyngis, and movements at the base of the tongue were produced. With extirpation experiments he found that eight dogs had lost the power of barking, on attempting which they uttered only a hoarse whine or made a sound.

Delavan⁷ in 1885 contributed a paper on the subject, collecting the facts recorded by Seguin, Krause and others.

In this article by Delavan is given the clinical history of a case which would appear to have a distinct bearing on the case we are recording. It is true that no necropsy was obtained in this case; nevertheless in the absence of any evidence of peripheral laryngeal disease the account of a left hemiparetic and associated unilateral cord paralysis apparently indicates that the latter was due to the cerebral lesion. A laryngoscopic examination in this case revealed much the same condition as was present in our case. The case of Delavan was one of the ordinary left hemiplegia in which the laryngeal and pharyngeal paralysis were especially well marked, and all the symptoms disappeared except the paralysis of the laryngeal abductors of the left side; these continued paralyzed up to the time of the publication of Delavan's article, a period of seven years. The laryngoscopic examinations in this case were recorded as showing the existence of a complete paralysis of the left vocal band, the position of which was apparently in the median line.

The report of Dr. Randall in our case states that some months before the operation the left vocal cord and arytenoid were immovable near the median line, and that some months after the operation the cord was atrophic-looking near the median line. Our case and

6. Krause (H.): *Archiv. für Anatomie und Physiologie, Physiologische Abtheilung*, 1883.

7. Delavan (D. B.): *Med. Record*, New York, Feb. 14, 1885.

that of Delavan, which seemed to be due to the cerebral lesion, have a striking similarity.

Garel,⁸ in 1886, reported a case in which the lesions were located in the inferior portion of the right precentral gyre and in the foot of the right third frontal convolution. The lesions were mainly cortical, invading only to a slight extent the subcortex.

Dr. Felix Semon, working with Sir Victor Horsley,⁹ found that excitation of the anterior portion of the lower end of the precentral convolution so affected the glottis as to bring about phonatory closure of the vocal cords.

In 1889 Semon and Horsley¹⁰ gave a condensed account of their experiments on the brains of a large number of monkeys, dogs, rabbits and cats, made with the view of determining, if possible, the existence of both adductor and abductor cortical centers for the vocal cords. In the first place it might be said that their results throw no light on the existence of an abductor center, although they give very definite information as to the existence of a center for adductor movements. These results, partly in their own language, were as follows:

There is in each cerebral hemisphere an area of bilateral representation of the adductor movements of the vocal cords, situated in the monkey just posterior to the lower end of the precentral sulcus at the base of the third frontal convolution, and in the carnivora in the precrucial and neighboring gyrus. This area has a focus of intensest representation in the anterior half of the foot of the ascending frontal convolution. Stimulation of this point produces complete bilateral adduction of the vocal cords, which lasts as long as the stimulation is continued. Should, however, the latter be unduly prolonged the "besoin de respirer" overcomes the influence of the artificial stimulus and evokes a momentary but powerful adductor movement of the vocal cords. Stimulation of the more peripheral parts of the area evokes less and less perfect adductions as we proceed from the focus outward, and when the extreme margin of the area is excited only what is known as the "cadaveric position" is assumed by the vocal cords.

The above description applies to the monkey, dog and rabbit. In the cat we have observed curiously different condi-

8. Garel: *Annales des Maladies de l'Oreille et du Larynx*. Tome xii, 1886, p. 218.

9. Semon (Felix) and Horsley (Sir Victor): *Phil. Trans. Royal Soc.*, clxlix, 1888.

10. The Central Motor Innervation of the Larynx, *Brit. Med. Jour.*, Dec. 21, 1889, p. 1383.

tions, for which, at present, we are not prepared to offer any explanation.

The extirpation experiments of Semon and Horsley show that destruction of the laryngeal center in one hemisphere did not cause adductor paralysis, but that in the same animal when the corresponding center of the other side was excited, just as completely bilateral an adduction of the cords was produced as if the opposite area were intact. They conclude—a matter of great importance in connection with the present case—that there is no such thing as unilateral paralysis of the vocal cord from lesion of the cerebral hemisphere. Semon and Horsley further found that prolonged excitation of the cerebral laryngeal center caused a true epilepsy of the vocal cords, gradually spreading to the face, neck, head and upper limbs. They conclude also that the epileptic cry is not to be attributed to excitation of the centers in the oblongata, but rather is a stage in the cortical discharge called an epileptic fit.

Whether it could be possible that in our case the Jacksonian spasm, which clearly involved the face and extremities of one side, began with a laryngeal spasm or was accompanied by such spasm, we have no means of proving; nor do the observations on the spasm when observed throw any particular light on this question.

While Semon and Horsley were not able to produce abduction of the vocal cords by their experiments on the cerebral cortex, they state that excitation of the accessory nucleus causes abduction and nothing else.

Risien Russell, in an experimental investigation on the influence of the cerebral cortex on the larynx,¹¹ obtained some valuable results with regard to the existence of an abductor center in the cortex. The experiments were undertaken with a view to determining the existence of an abductor center in the cerebrum of the dog, but were accompanied by other experiments on the brain of the cat, which, as in the case of Semon and Horsley, gave some curious and differing results.

In these experiments Risien Russell determined that the effects produced by excitation of the cerebral centers were practically irrespective of the condition of the peripheral apparatus. Like Semon and Horsley, he

11. The Influence of the Cerebral Cortex on the Larynx. Proceedings of the Royal Society of London, lviii, June 20, 1895, pp. 237-238.

could not obtain any evidence of the existence of a center for unilateral movements of the vocal cord. Dividing the recurrent laryngeal nerve of one side transversely, for instance, he found that he was able to produce the physiologic movement of the cord on the unaffected side equally by excitation of the cortical center in either hemisphere. He said:

The question of inhibition of antagonistic muscles by electric excitation of the cerebral cortex, on the lines adopted by Sherrington with regard to antagonistic muscles in other parts of the body, was tested by first dividing the adductor fibers in both recurrent laryngeal nerves, leaving the abductor fibers intact, and then exciting the adductor center with strong induced currents; but no evidence of inhibition of the abductor muscles was obtained.

The major part of the paper deals with the movements of the vocal cords which could be evoked on excitation of different foci in the cerebral cortex. It was found that both in the dog and cat there existed a focus, excitation of which resulted in adduction of the vocal cords, and another near to this, stimulation of which resulted in abduction of the cords. While in the cat it was possible to differentiate these movements without any preliminary measures being adopted, it was otherwise in the dog, for it was only after the adductor fibers of one recurrent laryngeal nerve had been divided transversely that it first became possible to evoke abduction of the vocal cords on excitation of the cortex, though in subsequent experiments it was sometimes possible to evoke this movement on excitation of the cortex of the dog without adopting this preliminary measure. The other effect on the cords, which it was as a rule found most difficult to differentiate from that of abduction, was acceleration of their movements.

In further exploring the cortex it was found that on the anterior composite gyrus, below the abductor center, there existed a focus, excitation of which resulted in what is described as a clonic adductor effect on the cords, in which the cords were first brought into a position of moderate adduction, and then there was added rapid short to-and-fro excursions.

On passing within the confines of Spencer's area for arrest of respiration, it was found that in the peripheral parts of this area there existed three foci, excitation of which affected the cords in different ways. The most anterior of these foci was responsible for arrest of the cords in adduction, i. e., in the expiratory stage of their excursions; excitation of the focus behind this, and corresponding, probably, to Horsley and Semon's abductor center in the cat, was followed by arrest of the cords in abduction, i. e., their inspiratory position; while the most posterior focus, which is situated at about the junction of the anterior composite and anterior Sylvian convolutions,

resulted in intensification combined with acceleration of the movements of the cords when stimulated. Excitation of Spencer's chief focus for arrest of respiration on the olfactory lobe resulted in arrest of the cords in the position they occupy during expiration in dogs, and in the position they occupy during inspiration in cats.

The summaries of the physiologic experiments of Krause, Semon and Horsley, and of Risien Russell, indicate that a unilateral irritative lesion of the cortical center for movements of the larynx should cause bilateral symptoms, and that a destructive lesion of this center on one side could produce bilateral paralysis of the vocal cords. Nevertheless, the clinical case of Delavan and the laryngeal findings both before and after operation in the present case point to the possibility of a destructive lesion of the cerebral center for laryngeal movements producing a persistent unilateral paralysis of the vocal cord on the side opposite to the lesion.

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HEMICRANIOSIS AND CURE OF BRAIN TUMOR BY OPERATION.*

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Under the name of *hemicraniose* Brissaud and Lereboullet¹ describe a condition observed by them in two cases—a hypertrophy localized exclusively to a half of the cranium including the face. The first patient had epileptic convulsions. The enlargement of the head was first noticed about the age of two months, and it was slowly progressive and without pain. The patient had no headache, although he complained of a sense of heaviness in the head. His vision was good. He had no vomiting. The interesting feature of the case was the frontoparietal exostosis. The prominence did not extend into the occipital region nor beyond the median line. There was also a supraorbital enlargement of bone, but the rest of the face was not affected. This was purely a clinical case.

The second patient, a woman, had cranial hemihypertrophy dating from childhood, also signs of brain tumor, the latter of two and a half years' duration. She had also exophthalmos. The hemihypertrophy was right-sided, slight in the superior and inferior maxillary bones, more pronounced in the frontal bone, and enormous in the frontoparietal region. The slope of the prominence extended to the other side of the cranium, so that the enlargement was not strictly confined to one-

* Read in the Section on Nervous and Mental Diseases of the American Medical Association, at the Fifty-eighth Annual Session, held at Atlantic City, June, 1907.

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1. *Revue Neurol.*, June 15, 1903, p. 537.

half of the skull. It was hard and painless. The necropsy showed that the entire inner portion of the right side of the dura was covered with tumors of different sizes (angiolithic sarcomata). The largest of these had made a depression for itself in the brain. The inner left side of the cranium was not abnormal, but on the right side of the calvarium the dura was adherent in places, the bone was irregular, and tumors were found within the dura.

In both these cases there was frontoparietal and supraorbital hyperostosis, and in one a slight implication of the face. All the tumors that were found in the second case had their origin in the dura. The authors suppose that the dura which in youth as internal periosteum had contributed to the progressive development of the hyperostosis, after the period of growth was passed, had its activity directed in another way, causing neoplastic proliferation. The unilaterality in both cases was absolute, they say, but the picture they present of the second patient shows very clearly that the slope of the prominence on the top of the head passed beyond the median line. The picture is the reverse of the hemiatrophy of the face of Romberg confined to the distribution of the fifth nerve. The hemicraniosis seems to be almost limited to the distribution of the ophthalmic branch. The dura is supposed by its outer surface to cause proliferation of the bone, and by its inner surface the formation of tumors. Although no operation was performed nor necropsy obtained in the first case, the convulsions suggested the possibility of intracranial tumor.

Parhon and Goldstein have reported a case of hemicraniosis in a woman whose only signs of intracranial disease were hemiplegia and mental depression. An exostosis the size of a walnut was in the right parietal region. When the skull was opened the bony tumor was found to protrude on the inner side of the skull, and it had caused a depression in a tumor of the dura beneath it. The tumor was the size of a small orange and involved the greater part of the motor zone and a part of the frontal lobe. It was circumscribed and distinct from the brain. The dura over it was adherent to the cranium. The calvarium was in general thicker on the side of the tumor. The microscopic examination showed that the new growth was a psammoma or angiolithic

sarcoma. The bony tumor was very compact and hard. The authors did not notice carefully whether any hyperostosis was present in the supraorbital and frontoparietal regions. Parhon believed a relation of cause and effect existed between the tumor of the bone and that of the brain, and Babès thinks there is no doubt of this, as the consistency showed that the bony tumor must be very old, and that it probably caused the psammoma by the constant irritation of the meninges. Babès, in discussing the case at a meeting, spoke of its importance, as it was the first time he had seen a tumor of one nature give origin to another of an entirely different nature. The case of Brissaud and Lereboullet and that of Parhon and Goldstein are the only cases of hemicraniosis with anatomopathologic examination in the literature, Parhon and Nadjede² assert, and these authors add a third case.

Their patient, a woman aged 63 years, had apoplexy and remained left hemiplegic, but the clinical history is very incomplete. At the necropsy the dura was found adherent to the anterior part of the calvarium, and in this area a bony prominence was detected the size of a nut in the middle and posterior part of the right frontal bone. There was no enlargement on the external surface of the skull. A tumor the size of the half of a small orange was found growing from the dura over the prefrontal lobe beneath the bony prominence. Microscopic examination showed that the growth was an angiolithic sarcoma or psammoma.

The authors confess ignorance concerning the etiology of this condition, why the proliferation of the cranium should occur, and especially in its anterior portion, as a result of the activity of the external portion of the dura, and why from the activity of its inner portion origin is given to psammomata. In the case of Brissaud and Lereboullet there were multiple tumors, but in the other two cases the tumors were solitary.

Whether this hemicraniosis is essentially different from exostosis of the upper part of the cranium, such as is described in the recent paper of von Eiselsberg,³ is, I think, open to question.

A case of endothelioma invading the skull, not unlike my first case, has recently been reported by Barling and

2. *Revue Neurol.*, Nov. 15, 1905, p. 1017.

3. *Archiv. für klin. Chirurgie*, vol. lxxxI, p. 1.

Leith.⁴ The bone over the center of the tumor in the brain for an area of about three-quarters of an inch was slightly thickened. Apparently hemicraniosis was not perceptible in this case. Endothelioma growing from the dura and deeply indenting the brain may exist, however, without causing thickening of the skull, and I have such a specimen in my collection.

In reporting two cases of hemicraniosis I may call attention to the fact that the notes of my first case were read at a society meeting in 1899,⁵ but the proceedings of this society are not published.

In both my cases the enlargement was on the left side, and in one case was strictly unilateral and in the other almost confined to one side, although it implicated the other side of the cranium to about the same extent as did the enlargement in the second case reported by Bressaud and Lereboullet. In both my cases an endothelioma was found growing from the dura and pushing the brain before it. In both operation was attempted, but in the first case the bone of the cranium was so implicated by the tumor, and hemorrhage on this account was so profuse that all attempt to remove the tumor had to be abandoned. The disappearance of nearly every symptom after the first operation when the skull was opened in the second case was the result of decompression, and relief was obtained for several months. I think it a fortunate thing that the dura was not incised at the first operation, as the tumor would then have been discovered and its removal attempted. The time given to the patient for complete recovery after the first operation made him more vigorous to endure a second, and the tumor was removed later with comparatively little danger to the patient. The cranium in my second case was very dense, but was not infiltrated by tumor; had it been, as in the first case, the result of operation would have been grave.

Disappearance of all symptoms after removal of a brain tumor is exceedingly rare. The most successful case in my own experience previous to the one I now report was that of a parietal tumor reported by Dr. C. K. Mills, Dr. W. W. Keen and myself. This patient I saw in consultation with Dr. Mills a number of times, and

4. *Lancet*, Aug. 4, 1906, p. 282.

5. The Section on General Medicine of the College of Physicians of Philadelphia.

improvement lasted about four years after the removal of the tumor, but recurrence then occurred and a second operation proved fatal. The hemiplegia in this patient never disappeared entirely.

In one of the cases⁶ I now report almost every symptom disappeared since the second operation. The patient's speech is very nearly or fully normal, if he does not talk too rapidly; he has no hemiplegia, no optic neuritis, no disturbance of writing, little or no paresthesia of the hand, little or no abnormality of gait, and no exaggeration of the patellar reflexes.

In a recent discussion on cerebral and spinal tumors Krause⁷ remarked that true recovery from brain tumor after operation is always very rare. Schultze reported that in 19 operations performed on 97 cerebral tumors only once recovery occurred, lasting a few years after the operation, and the case was one of cerebellar tumor. Oppenheim has had 27 cases of tumor of the brain with operation since 1903. Of these 3 were with cure, 6 with temporary improvement, 15 with fatal termination. Bruns says he has never had entirely satisfactory results in operation on brain tumors, and von Monakow has had only one successful case, and in this the tumor was in the motor region. Horsley, at the meeting of the British Medical Association in Toronto, in 1906, exhibited the photographs of a patient from whom a cerebellar tumor had been very successfully removed some time ago.

A case of brain tumor with disappearance of the

6. Note in correcting the proof Oct. 31, 1907: During the past few months the swelling on the head directly in front of the opening in the skull increased markedly, and on Oct. 23, 1907, a convulsion confined to the right upper limb and right side of the face occurred. It is not known whether the right lower limb was affected or not. An operation was decided on. The swelling was well defined superficially, but was adherent to the bone. The bone beneath it was about one-half inch in thickness and softer than normal bone. It was easily cut with a knife, so that the outer portion was removed with the tumor. The latter was firm and fibrous in appearance. A large tumor was found beneath the dura. It was very soft, and was entirely removed with a spoon. The cavity was lined by the cerebral cortex, the convolutions of which could be seen at the upper margin. It measured $1\frac{1}{4}$ inches in depth, $3\frac{1}{2}$ inches from before backward, and 3 inches from above downward. The piece of dura from which the tumor had grown was found and removed. Microscopic examination showed the tumor to be an endothelioma. The operation was performed by Dr. C. H. Frazier. The patient made a rapid recovery, was not in danger at any time, had no symptoms of cerebral disease, and left the hospital ten days after the operation.

7. Monats. f. Psych. u. Neurol., January, 1907, p. 87.

symptoms after operation has been reported by Oppenheim and Krause.⁸ The patient had right hemihypoesthesia, hemiataxia, hemiparesis, hemianopsia and headache. A tumor was removed from the occipital lobe. Two months later the patient had merely some difficulty in reading and the visual field of the right eye was a little contracted.

A still more brilliant case of cure of brain tumor by operation is one described by Oppenheim and Borchardt,⁹ in which a tumor the size of an egg was removed from the cerebello-pontile angle. The symptoms, which were very grave, gradually disappeared after the operation, and eight months later the patient could be pronounced cured.

In both my cases a history of head trauma was obtained; in the first it occurred about eight years before the enlargement of the head was noticed; in the second only a short time previously. It is not improbable that these injuries were at least the predisposing cause of the bony proliferation.

The presence of tumor now in five cases of hemi-craniosis with operation or necropsy, and probably also in the clinical case of Brissaud and Lereboullet, should make one fear that any pronounced local thickening of the cranium is likely to be associated with a tumor growing from the dura, and operation should be attempted as soon as cerebral symptoms become manifest. It may be possible to remove the tumor in this way while it is yet small. Fortunately, this local thickening of bone is usually over or near the motor area of the brain, so that the symptom-complex is distinct, and the tumor growing from the dura is not likely to infiltrate the brain. Operations on these dural tumors are especially serious because of the severe hemorrhage, and they might be more successful if done in two stages.

The absence of all those symptoms which are frequently attributed to lesions of the left frontal lobe, in my second case after a large tumor had been removed from this lobe, is striking. The patient is a man engaged in a large and important business, and is as capable of performing his duties now as at any previous time. There is no failure of judgment, intelligence, power of attention, concentration of thought, memory,

8. Berlin. klin. Wochschr., Dec. 17, 1906.

9. Berlin. klin. Wochschr., July 15, 1907, p. 875.

or of any of the intellectual functions. The tumor acted by pressure, not by infiltration.

To what are we to attribute the transitory aphasia, agraphia and hemiplegia following the second operation? Pressure on Broca's area explains the first very readily, but if we accept Marie's views we must assume that pressure was exerted on the lenticular nucleus, which was remote from the seat of operation. It is difficult to attribute the disturbance of speech and writing that developed shortly before the second operation to pressure on the lenticular nucleus, because the motor cortex or subcortical motor fibers were between the tumor and the lenticular nucleus, and the hemiparesis was not so intense as the speech and graphic disturbances, indicating that the centers or fibers of motion for the limbs were less implicated than those of speech and writing.

The first case is as follows:

CASE 1.—J. T., aged 47, male, came to my service at the Polyclinic Hospital in the beginning of 1899.

History.—His wife had not had any miscarriages or still-born children, and no history of venereal disease was obtained. He had taken iodids without benefit.

The following notes were read by me before the Section on General Medicine of the College of Physicians of Philadelphia, May, 1899, and the title of the paper appears in the Transactions of the College for that year, p. 191. It was my intention to publish promptly these notes in full in association with the very complete report of Dr. T. S. Kirkbride, Jr., which follows, but the sad death of Dr. Kirkbride delayed the publication of them and made me lose interest in the case.

At the last meeting of the section (April, 1899) I presented a patient who was partially paralyzed on the right side of the body and had exostosis of the cranium. My object in doing this was to exhibit the man before operation was performed. The patient said that his right hemiparesis had developed after influenza; that he had been very much better until he had had another attack of influenza, when his paresis increased. I found later that his statements were not reliable, and that his memory was poor. His wife was not very intelligent and was not of much service in obtaining a correct clinical history. An intimate friend of the patient gave me the important details:

About sixteen years ago the man struck his forehead in descending a staircase. The severity of the blow can not be determined, but it was sufficient for the blow to be remembered after sixteen years. About eight years later the patient noticed that his head was becoming enlarged, and it is probable that the head had increased in size before the man's attention was

called to it. About three or four years ago he began to have paresthesia in his right toes, and he examined these parts frequently in order to detect a sore of some kind, but none was found. The symptom was probably indicative of the beginning cortical lesion. At the same time his right leg began to grow weak, and after a year or two he felt similar paresthesia and weakness in the right upper limb. A few months ago he had his first and only attack of eleptiform convulsions. His wife awoke and found him in general convulsions and unconscious. One day in the summer of 1898 he was aphasic for ten minutes, and was unable to say a word, but was entirely conscious. He had had similar attacks previously, less pronounced. This temporary aphasia was probably a sign of pressure affecting the speech area, and the convulsions were indicative of cortical irritation. He had not suffered to any extent from headache, nausea or vertigo. About a week before the operation, however, he began to have nausea and considerable headache.

Examination.—When he was presented at the last meeting of the section he was hemiparetic on the right side, but the face was not distinctly involved, objective sensation was intact, the patellar reflexes were exaggerated, especially that on the right side. The memory was poor, the station was good, the patient was partially aphasic. He knew the word he wished to say but was often unable to utter it, and was obliged to select other words than those which best expressed his meaning; to this extent he was paraphasic, but he had no true paraphasia, no jargon aphasia of the English writers. He did not mix the parts of speech without regard to the proper position of nouns and verbs; all he said was perfectly intelligible, and this is comprehensible from the fact that the necropsy showed that the speech area was only involved by pressure. The exostosis of the cranium was very striking. It began above the line of the hat brim on the left side in the area of the left temple, extended backward over the left Rolandic area and crossed the median line. The pupils were equal and the reaction to light and accommodation was good. Dr. Hausell reported that the man had bilateral optic neuritis going on to atrophy with limitation of the visual fields for form and color, and, later he stated that the disease of the optic nerves appeared to be a chronic rather than an acute process.

Summary of Case.—In brief, the important features of the case were these: A blow on the head sixteen years ago; enlargement of the left side and upper part of the skull extending over the left Rolandic area, first noticed by the patient about eight years ago; paresthesia, and weakness in the right lower limb developing three or four years ago; a similar condition appearing in the right upper limb a year or two later; symptoms of aphasia, failure of memory, epileptiform convulsions, exaggerated patellar reflexes, bilateral optic neuritis of a chronic type, and later nausea and severe headache. The

case seemed to me one entirely suitable for operation. Some disturbance of the sense of position in the right foot, the aphasic symptoms and the convulsions indicated a cortical lesion of some kind and the external bony enlargement suggested the possibility of a similar growth on the inside of the skull pressing on the leg center, for the cortical lesion evidently began in the leg center. The right-sided paresis was not complete paralysis, the intellect was not much impaired, and the general health was excellent. The symptoms were undoubtedly those of intracranial pressure.

In addressing the students of the Polyclinic Hospital before the operation was begun I said that we could not be positive that a bony growth would be found on the interior of the skull exactly similar to that on the outside; but that the existence of such a growth was quite possible and would explain the symptoms; that the most we could say was that the man had signs of focal intracranial pressure in the upper part of the left Rolandic area and that operation was demanded.

Operation.—Dr. Max J. Stern and Dr. John B. Roberts examined the patient and an operation was performed, April 15, 1899, by Dr. Stern, assisted by Dr. Roberts. A large bone flap was raised in the region of the thickened bone, but the hemorrhage was so profuse that the operation was stopped without any further attempt being made to remove the tumor, and the patient died about two hours later from loss of blood.

AUTOPSY.

The necropsy was performed by the late Dr. Kirkbride, and the following notes were read by him in May, 1899. (Examination of head only. A complete autopsy was refused by the relatives of the patient.):

The scalp of the vertex, on removal of the dressings covering the head, is seen to have been turned back at the operation in a large flap with the base posteriorly. The under surface of the flap is, in the left parietal region, considerably lacerated, evidently owing to the breaking up of firm adhesions between the pericranium and the bones of the skull. In the same region the tissues of the scalp are indurated, there being marked increase in the fibrous tissue between the bundles of which there are here and there small spicules of bone. The vault of the cranium has been removed at the operation in a large oval area about 9x12 cm. in size, with the center slightly to the left of the middle line. The portions of bone removed at the operation vary in size from 0.9 to 2.5 cm.; the largest is 10.5 cm. long by 4 cm. broad. On the outer surface of this piece there are irregular, partly nodular exostoses, some firm and closely adherent to the underlying bone, others honey-combed and fri-

able. Shreds of pericranium adhered to the central portion of the outer surface of this fragment. The inner surface is in its central portion distinctly convex and is uneven, with shreds of dura still adhering. Where the dura has been torn away the bone has a honey-combed appearance. On sawing through the fragment the cut surface shows an almost uniformly indurated bone, the location of the diploë appearing simply as an indistinct band of darker color and less firm consistence between the more compact outer and inner tables which are greatly thickened. The smaller fragments of excised bone resemble the one just described, but are in general thinner and with less evidence of adherent pericranium and dura. On the inner surface of one of these smaller pieces there is a nodular exostosis measuring 4x6 mm. and projecting 3 mm. from the surface.

The remaining portion of the vault of the cranium removed at the autopsy presents no abnormalities, the impressiones digitatæ are not deep.

The dura is throughout somewhat more than normally adherent to the bone. It presents an uneven, lacerated appearance in an irregular area measuring 5.5x7.5 cm. located over the upper left motor region and extending 1 to 2 cm. beyond the middle line on the right (seat of operation). The membrane is here of a dark red color and appears much thickened. In the surrounding portions of the dura there are scattered spots of bright red color (hemorrhage) and throughout the membrane is moderately thickened. Over the right hemisphere the dura is not adherent except along the central portion of the longitudinal fissure, and here it is easily detached. In the adherent portion are several small bony formations. The dura of the left hemisphere appears to be firmly adherent over a considerable area and is therefore left in place. On the surface of the dura about 3 cm. in advance of the middle portion of the Rolandic fissure is a nodule of bone measuring 1 cm. in length and 0.5 cm. in width. The cerebral cortex is slightly depressed by this bony formation. On the left surface of the falx immediately below the collection of small bony growths mentioned in connection with the right hemisphere, there is a thin lamina of bone 1 cm. long resting against the median surface of the left hemisphere. Directly posterior to this, also on the left surface of the falx, is a similar bony plate measuring about 2 cm. in length and 1 cm. in width. The longitudinal sinus is much narrowed where it is included in the region of thickened dura; it is patulous, however, and contains no thrombi. The removal of the brain shows the apices of the temporal lobes adherent to the dura; in the same region the impressiones digitatæ are unusually deep, otherwise the base of the skull is normal.

The brain and the posterior half of the left eye were removed and placed in 7 per cent. formol.

The brain preserved in formol: The right hemisphere, except

for the adhesion of the dura already noted, appears normal. Over the left hemisphere there is an area of marked depression 4 cm. in width and 3 cm. anteroposteriorly, which in its posterior portion encroaches on the upper part of the Rolandic fissure and which extends on the right as far as to the longitudinal fissure. The dura appears in this area to be firmly adherent to the cerebral cortex. The portion of dura apparently adherent to the cortex is more extensive than the area of depression, measuring 7 cm. anteroposteriorly by 5 cm. laterally. Careful examination, however, proves that the apparent adhesion to the cortex is in reality the broad pedicle of a tumor arising from the dura itself and projecting far into the substance of the brain which has been pushed aside and compressed so as to form a deep socket in which the tumor lies. The pedicle measures 7 cm. anteroposteriorly by 5 cm.

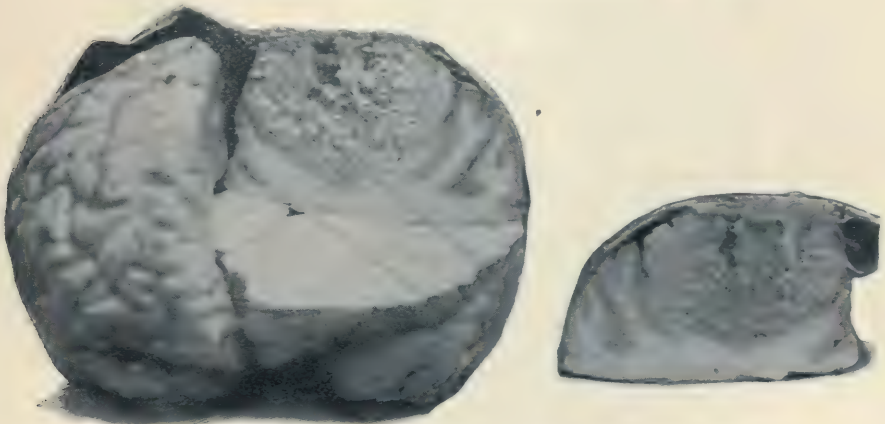


Fig. 1.—Case 1. Showing the tumor (endothelioma) in the brain

laterally. On turning back the dura the tumor is seen to project on the left fully 1 cm. beyond the margin of the pedicle and it here appears as a convex grayish, firm gyrus-like mass beneath the pia mater; indeed, the resemblance to the neighboring convolutions was so marked that this portion of the tumor was at first mistaken for a convolution infiltrated by extensions of the tumor. Anteriorly and posteriorly the tumor projects but little beyond its attachment to the dura. On turning back the falx, a portion of the tumor covered only by the pia is visible on the median aspect of the hemisphere. This exposed surface of the tumor is irregularly semicircular in shape, measuring anteroposteriorly 4.5 cm. and extending downward in its broadest part fully 2 cm. below the circumference of the hemisphere. Everywhere at the margins of the socket in which the tumor lies the pia mater can be seen dipping down between the tumor and the cortex. (Figs. 1 and 2.)

Position of the tumor: The fissure of Rolando, if not distorted, would in its upper portion pass through the center of the tumor. It occupies a little more than one-half of this fissure. The division of the left hemisphere in a frontal plane through the center of the tumor shows the latter to be much larger than appears from the surface. It is an oval mass, flattened on its upper surface, measuring 6 cm. transversely, and projecting downward into the hemisphere fully 4 cm. from the level of the dura. This measurement does not, however, represent the total encroachment on the brain substance, for, as already mentioned, the upper surface of the dura (i. e., of

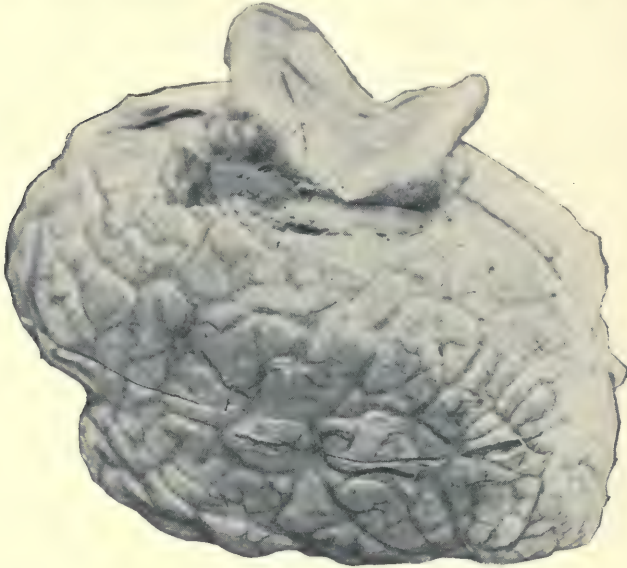


Fig. 2.—Case 1. Showing position of the tumor attached to the dura.

the tumor) is in this area concave, adapting itself to the convex inner surface of the greatly thickened skull. The socket of brain substance in which the tumor rests is lined by pia mater, beneath which a narrow band of gray matter indicates the position of the greatly compressed cortex which the tumor has evidently pushed before it in following by its growth the lines of least resistance. Beneath the center of the tumor, where the compression has evidently been most marked, the gray matter has almost entirely disappeared. On the median aspect of the tumor the compressed cortex tapers to a sharp edge, above which the tumor lies directly beneath the pia mater as already noted. The tumor itself appears on the cut surface of a grayish-white color. Its consistence is moderately soft ex-

cept where in the upper central portion it contains bony particles. In this part the surface appears more compact and evenly granular, while below and at the margins projecting beyond the pedicle its formation is looser and more sponge-like. The surface is rather dry and mealy.

The uppermost part of the ascending parietal convolution appears to have been compressed by the tumor and is diminished in size. The second and third frontal convolutions are not involved and the area of Broca seems also to be intact. The median and middle portion of the left hemisphere is convex (on its median aspect) by the pressure of the tumor, while the median and middle portion of the right hemisphere is concave and adjusted to the convexity of the left. The gyrus fornicatus of the left hemisphere extends almost completely across the free surface of the corpus callosum. The posterior horn of the right ventricle is two or three times larger than that of the left. The aqueduct of Sylvius is very slightly enlarged. The right side of the optic chiasm is much flattened from below upward with the concavity on the lower surface. The left side is normal in shape. The right optic nerve near the chiasm is possibly one-fourth smaller than the left.

The following notes on the ocular findings are from Dr. E. A. Shumway:

The posterior portion of the left eye submitted for examination was embedded in celloidin and cut in sections parallel with its horizontal meridian. Transverse sections were also made through the optic nerve posterior to its entrance into the globe. The latter show very slight changes in this portion of the nerve, the intervaginal space being slightly dilated and the endothelial cells lining the processes of the arachnoid being increased in number. The nerve fibers have retained their myelin sheaths and show no evidences of atrophy. The meridional sections show a swelling of the nerve head, within the lamina cribrosa, which reaches an elevation of 1.09 mm. above the level of the chorioid. The retina at the optic nerve entrance is detached, somewhat folded, and is separated from the underlying chorioid by a layer of thickened nerve fibers. The non-medullated nerve fibers are swollen, show well-marked varicosities, and are separated from each other, evidently as the result of pre-existing edema. This condition of the fibers can be followed for some distance in the neighboring parts of the retina. There is a moderate infiltration with cellular elements, which are assuming a spindle shape, and indicate the beginning of a connective tissue formation.

The retina is detached throughout the specimen, and the layer of rods and cones shows postmortem changes. The nerve fiber layer is thickened, edematous, and the fibers of Müller show marked hypertrophy in places, producing irregular elevations

on the outer surface of the membrane. No hemorrhage is present in either retina or nerve.

The chorioid is very densely pigmented. At a point 5 mm. to the nasal side of the optic nerve entrance is a deeply pigmented growth, which measures 6 mm. in its greatest vertical, and 4 mm. in its greatest horizontal diameters, and 0.9 mm. thick. It is composed largely of pigment cells which are evidently proliferated stroma cells, but shows also a large number of non-pigmented and round spindle cells with large, deeply staining nuclei, closely packed between the chorioidal vessels. The growth has apparently sprung from the posterior layer of the chorioid, but the cells have infiltrated the anterior layers, containing the smaller capillaries. The pigment cells of the retina pass over its surface undisturbed, and the retina shows no evidence of inflammatory reaction. In the neighborhood of the growth the chorioidal vessels are widely distended, and both the vessels and the chorioidal stroma show a slight leucocytic infiltration. The growth is therefore to be diagnosed as a melanosarcoma of the chorioid.

Diagnosis: Papillitis with swelling of the nerve head 1 mm. above the chorioidal level; moderate circumpapillary retinitis; beginning melanosarcoma of the chorioid.

This finding of a melanosarcoma of the chorioid by Dr. Shumway in association with an endothelioma of the cerebral dura is interesting.

CASE 2.—A. B., male, aged 42, was referred to me June 28, 1906, by Dr. H. C. Westervelt, when the following notes were made. He had been under the care of Dr. J. I. Johnston.

Chief Complaint.—An enlargement in the left parietal region; right-sided weakness, staggering to some extent; some motor aphasia, with slow indistinct speech, possibly sensory aphasia.

History.—Six or seven months ago he observed a gradually increasing swelling in his cranium over and anterior to the left parietal region. This was accompanied by severe left-sided headache. Several weeks later he found himself unable to articulate correctly and stammered and mouthed his words indistinctly when speaking. There was no weakness in the arms or legs, but he has felt numb in his right arm and leg, and occasionally staggers rather badly when walking. Otherwise there have been no sensory disturbances. His sight, so far as he knows, is good, but he can not read well, as gradually the lines appear to waver on the page. His memory is not nearly so good as it was; when speaking he has definitely in mind what he wants to say, but is slow in getting words to express himself, and then speaks hesitatingly and with apparently great effort. His voice, he believes, has grown weaker. He has no defect in hearing. About two months ago, he was at Mt. Clemens for five weeks, where he underwent treatment. After he returned, he had hypodermic injections of bichlorid of mercury followed by inunctions. With this treatment he

thinks the tumor in his head lessened in size and that his speech and memory improved to some extent. But during the past four weeks he thinks they have grown worse again.

Examination.—His bowels are constipated, appetite is poor, tongue coated, and he has lost 15 pounds since January. He sleeps well. Occasionally there is some pain over the af-

A

Dr. Wm T. Spiller, *July 26th 1906*

Dear Doctor:

I intend to consult Dr. Randall in a few days. I shall, therefore, be obliged, if you will kindly write to the doctor, as you proposed.

Very truly,

B

It is so hard for me to write, that I know you will ~~excuse~~
I guess you ~~too~~ had better not ~~not~~ get ready to come ~~there~~ here. I may go to the Philadelp^h in a week or a week.

Fig. 3.—Case 2. A, writing 19 days after the first operation. The man could not write at all for some time before the operation. B, writing 14 days before the second operation.

ected part of his head. No tremors or convulsive movements have occurred in face, arms, or legs. On admission to the hospital, temperature, pulse and respiration are normal. His wife says that in the beginning of the present disturbance he staggered in walking like a drunken man, and at times (lasting 15 or 20 minutes) seemed utterly unable to speak. Physically, he is well developed, somewhat poorly nourished, flabby; with

very moderate panniculus adiposus. On the left side of his cranium, extending forward from above and behind his left ear there is an extensive, gradual swelling, hard and not painful to touch, producing slight asymmetry of his head.

Eye examination by Dr. de Schweinitz, June 30, 1906: Pupillary reactions are normal. Extraocular muscles are normal. In extreme outward rotation on each side there is very slight oscillation of the eyeball of each eye. Fundus shows no evidence of optic neuritis. There is slight over-fullness of the retinal vessels. Color perception normal on both sides.

On looking at the ceiling the wrinkles in the right side of the forehead are not quite so distinct as those in the left. In closing the eyes the furrows about the right eye are not so deep as those about the left, but there is no weakness of the orbicularis palpebrarum. When the face is at rest the right corner of the mouth droops a little, and the right nasolabial fold is not quite so distinct. In showing his teeth the right nasolabial fold is not so deep as the left. He can not draw up the right corner of the mouth separately nearly so well as the left. Speech is somewhat thick, and while he speaks without any great interruption, he has evidently at times difficulty in getting the word he wants. His memory is poor. There is no history of diplopia. The tongue deviates to the right when protruded. The extraocular muscles seem to be very good. Pupils are not quite equal, the right being a little larger than the left. In looking at a near object the pupil dilates in each eye; in looking into space the pupil of each eye at first contracts distinctly and then dilates. Reaction to light is prompt in each eye, as is also contraction in convergence. A little involuntary jerking occurs occasionally on the left side in the lower part of the face. (Dr. Jones says he has had this facial tic since he has known him, 23 years.)

The swelling above and in front of the left ear is very distinct and large. He winces a little when the tumor is palpated. He has had a little vomiting at times, probably due to iodids. He has neither word blindness nor word deafness. Sensations for touch and pain are normal in both sides of the face.

The grasp of the right hand is fair, but not so good as that of the left. He has been a right-handed man, and he has noticed that the right hand has become a little weak. Sensations for touch and pain are normal in each hand. Stereognostic perception is normal in each hand. Biceps and triceps reflexes are distinct on each side, a little prompter on the right. Sense of position is normal in the fingers of each hand. Resistance to passive movement is a little weaker in the right upper limb. Patellar reflex is prompt on each side, and exaggerated a little on the right. Standing erect with eyes open he sways a little, and the sway is not distinctly increased by closing the eyes. The gait is very good with eyes open or closed. Slight indication of ankle clonus is detected on both

sides, but no Babinski. Sensations for touch and pain are normal in each foot. Achilles jerk is prompt on each side, and about equal. He can not take solids because of bloody purging.

July 3, 1906: He was observed stumbling again to-day, as he did a few days ago. He is in a very excitable state and is a little unstrung. He is now eager for operation to rid him of his symptoms. Speech is, if anything, more indistinct than before. Weakness in the right upper and lower limbs is not more marked, but he sometimes carries his right arm while walking, as if he were afraid of hurting it.

July 5, 1906: He was asked to write a letter, but after writing a few words stopped and was not able to go on. He is getting very nervous and has the greatest difficulty in articulating.

July 7, 1906: Transferred to Dr. Frazier's service for operation.

Operation.—A section of bone comprising the outline of the tumor mass was removed by means of trephines and chisels. It was greatly thickened. The dura was found somewhat thickened, but was not opened, as the abnormal thickness of bone seemed to be sufficient to account for the symptoms. The man made an excellent recovery from the ether and was only slightly nauseated. Microscopic examination of the bone removed showed no appearance of tumor cells.

Subsequent History.—The patient made almost a complete recovery, attended to his business as formerly, and seemed well, but had some return of symptoms a few months later.

Sept. 4, 1906: He yawns much. The site of the operation is depressed and tender to touch. His right hand feels as though it were asleep and is numb. His speech is a little thicker than at his last examination, but he has no aphasia. The grasp of each hand is good. It requires a little effort for him to draw up the right corner of the mouth.

Oct. 23, 1906: His speech has become affected, is slow, and a little thick. He has much headache and is weaker than he was, but is not dizzy.

Nov. 10, 1906: The speech is much worse than it was ten days ago. He can say short sentences, but so soon as he attempts to tell anything, he becomes much confused and many words he can not speak. If he speaks slowly and is not excited he can make himself understood. He can not write correctly, as shown by the letters he attempts to write to his wife. He has had severe headache for the past few mornings. The grasp of each hand is fair. He has had difficulty in walking down stairs during the past two or three weeks. Resistance to passive movements is good in the lower limbs. The patellar reflex is exaggerated on each side, more so on the right side. The mental condition is good. It was evident that some lesion was developing at the site of the former operation, and fearing tumor, I referred the patient again to Dr. Frazier.

Second Operation.—An operation was performed by Dr. Frazier November 17. The old skin flap was opened. Bleeding was very profuse. The dura was cut and a tumor was found in the center of the opening. It was shelled out, although in so doing the forward part, which extended beyond the original opening, was broken, and was taken out in fragments. (Fig. 4.) Its position must have been above Broca's area and in front of the central fissure.

Result.—The patient was completely paralyzed on the right side and completely motor aphasic as a result of the operation. He began to recover the power of his right limbs within a few days, and within four or five days he had recovered almost as much power as he had before the operation, when he showed slight hemiparesis. After a few days he could say "Yes," "No," and one or two other short words. Gradually he became able to add a few more words, and those he said were usually distinct. On November 23 he was able to say a number of words in repeating after the examiner, and a few spontan-

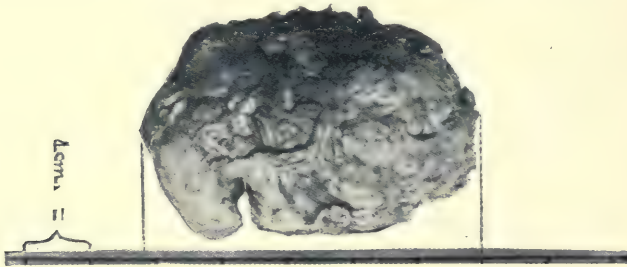


Fig. 4.—Case 2. Tumor removed.

eously, but he could not put words together. He occasionally said two or three words with great difficulty. He was perfectly intelligent and not at all word deaf. He showed no intellectual defect. He could not write except that he managed to write, "Look at them." He read and obeyed five simple commands, when written, such as: "Put your right hand on your ear." He was not ataxic, and recognized words in the newspaper (Fig. 5).

Dec. 1, 1906: There is some weakness of the right muscles of mastication, the tongue deviates a little to the right, the right side of the face in the lower distribution is a little weak, Motor speech is still disturbed but is much better. He can say sentences and make himself understood, but many words he can not get and his sentences contain only a few words. The grasp of the right hand is hardly weaker than that of the left and is about normal. He picks up small objects with his right hand without difficulty. The sense of position and stereognostic perception are normal. The power in each lower limb is normal. The patellar reflex is prompt on each side, a little

more so on the right. Two or three days after the operation he was able to communicate with his wife by pointing to letters she showed him so as to spell words.

Dec. 18, 1906: The speech is greatly improved and only occasionally he has difficulty in getting a word, but his words are sometimes indistinct. His mentality has in no way been impaired. There is still a little weakness of the lower part

A

Stetter
 who wife
 run

B Philadelphia 11/25/07

Dr W G Spiller,

This is a specimen
 of my handwriting
 yours

Fig. 5.—Case 2. A, writing done with the left hand 3 or 4 days after the second operation. The right hand could not be used in writing. B, writing on eighth day after the second operation.

of the right side of the face. He complains of some numbness of the right thumb and of the right upper lip and right gum. There is a little swelling over the seat of operation, but this is not distinctly painful. The grasp of the right hand is excellent. He has no disturbance of stereognostic perception in the right hand. Both patellar reflexes are prompt, the right more so.

The opening in the skull according to Anderson-Machin lines is $\frac{3}{4}$ inch behind and $2\frac{1}{4}$ inches in front of the fissure of Rolando. The opening at its greatest width is 3 inches (Fig. 6).

Dr. de Schweinitz reported Dec. 19, 1906: "The slight edema of the nerve head, the so-called edematous optic neuritis, which I observed when last I saw the patient, has entirely disappeared and vision is absolutely normal. I made some suggestions in regard to a change in the use of his glasses, which I hope will be of service. There are no muscular anomalies, and the field of vision is normal."

Dr. de Schweinitz' previous report, without date, but made after the patient had entered the hospital for the second operation, and probably just after the operation, to which he refers, is as follows: "V. 4/4. No diplopia; some nystagmus in outward rotation; slight but distinct optic neuritis, more in the right eye; veins very full; temporal edges not yet involved."

Jan. 22, 1907: Speech is still thick, but the man says all that he wishes to. Both patellar reflexes are a little prompter than normal. The tongue is protruded straight. The lower



Fig. 6.—Case 2. Tracing of photograph showing the position of the bone flap.

part of the right side of the face is a trifle weak. The grasp of each hand is excellent. He feels splendidly. The numbness in the right hand has disappeared except in the thumb. He has no headache and no vertigo. Stereognostic perception is normal. He says that a year and a half ago he received a severe blow against a beam at the region where the enlargement of the head occurred. This information was not obtained previously. He was nearly unconscious from this. If he speaks slowly he speaks distinctly.

March 28, 1907: He has been feeling so well he did not think it necessary to see me. Very slight weakness of right lower part of face may be detected. Grasp of the right hand is normal. The patellar reflex is about normal on each side. Gait is normal. A note from Dr. de Schweinitz of this date is as follows: "Vision is normal and there are no ophthalmoscopic lesions in the eye grounds; perhaps a little over-fullness of the retinal veins. Pupil reflex normal; no diplopia. No change in the right field; moderate concentric contraction of left field. Slight reduction of light sense (6/10)."

ARTERIOSCLEROSIS OF THE CENTRAL NERVOUS SYSTEM, WITH A REPORT OF THREE CASES.

BY HENRY S. HUTCHINSON, M.D.¹

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CASE I.—Devenny; spastic paraplegia; admitted January 24, 1906, in semistuporous condition, to the service of Dr. Spiller.

Physical Examination. An aged woman, sparse white hair; chest apparently clear, even hyperresonant to percussion. No heart murmurs. Arteries markedly sclerosed; pulse feeble; abdomen board-like.

Extremities. The arms are flexed at elbows and crossed over the chest; muscles are spastic; the arms can be straightened when some force is used. Legs: Thighs flexed on abdomen and legs on thighs, especially the left leg, which lies directly for eight inches below the knee upon the thigh. On account of the spasticity, the reflexes cannot be obtained.

¹ Dr. Hutchinson was engaged in preparing this paper in the Laboratory of Neuropathology during his fourth year as a medical student at the University of Pennsylvania, and completed it shortly before his death. It is published as written by him.—WILLIAM G. SPILLER, M.D.

Only complaint is of being cold. Does not like to be disturbed for meals or for medicine.

February 1, 1906. Notes by Dr. Weisenburg: Patient's mentality is poor. Says she is worried because she does not feel well; she has no pain anywhere; seems to have bilateral cataract. Biceps and triceps reflexes are present and prompt. Has incontinence of urine and feces; no Babinski; sensation apparently normal.

Notes by Dr. Spiller: Both upper and lower limbs are markedly contracted in flexion and wasted, as is the whole body. The upper limbs can be forcibly extended at all parts, including fingers, but the limbs are very stiff. Mentally the patient is so feeble that no history can be obtained. Biceps tendon reflex is very prompt on each side. Lower limbs cannot be forcibly extended as the contractures are so great. It is impossible to determine whether patellar tendon reflexes and Achilles jerks are exaggerated or not, because of the rigidity of the lower limbs. Pricking feet with pin causes pain. Diagnosis: Senile changes of the central nervous system.

February 17, 1906. Patient is in a stuporous condition. Hands and feet and face are cold. Pulse rapid and weak. There has been no bowel movement, in spite of enemata and medicine. Died February 18, 1906.

Microscopic examination of cord and brain showed the following:

Lumbar Region. There are distinct degenerative changes in the nerve cells of the anterior horns, these changes consisting in a breaking up of the chromophilic elements in the centre of the cells, with occasionally peripherally placed nuclei. The cells are heavily pigmented. The small vessels, especially in the posterior columns, show greatly thickened walls; and numerous bands of neuroglia are found within the cord substance, so that there is a considerable degree of diffuse sclerosis. There is no distinct degeneration of nerve fibers, either in the cord itself or in the anterior and posterior roots.

Thoracic Region. The thickening of the small bloodvessels within the cord is more pronounced here than in the lumbar region, and the sections are like those of certain cases of paralysis agitans. Sections stained by the Mallory connective-tissue method demonstrate these thickened and multiplied bloodvessels very well; and this stain also colors the amyloid bodies, which are seen in very large numbers, especially at the periphery of the cord.

Cervical Region. The cells of the anterior horns are altered in the same way as those of the lumbar region. Sections stained by the Marchi method, both of the lumbar and cervical region, show only a few scattered black dots, which are not indicative of degeneration.

Medulla oblongata shows nothing abnormal beyond the presence of numerous amyloid bodies.

Paracentral Lobules. The cells of Betz in both paracentral lobules show some alteration, consisting in central chromatolysis, displacement of the nucleus, and intense pigmentation. Even the small cells of the cortex are deeply pigmented. The small vessels are also much thickened. Amyloid bodies are exceedingly numerous at the periphery of the cortex.

Optic Chiasm and Optic Nerves. No degenerations are present in the optic nerves. The septa of the chiasm are greatly thickened, as are also the walls of the bloodvessels; and in the optic nerves a marked increase in neuroglia tissue is present. At the sides of the chiasm, numerous amyloid bodies are seen.

Olfactory Tracts and Bulbs. Along the course of the olfactory tract amyloid bodies in enormous number are seen, scattered between the fibers, and diminishing in number as the olfactory bulb is reached. The Mallory connective-tissue stain colors these bodies a deep blue.

CASE II.—Johnson, an old woman, unable to talk, was admitted to the Philadelphia General Hospital, service of Dr. Spiller, August 25, 1905. Notes by Dr. Weisenburg:

Voice weak, uncertain; speech understood with difficulty. Pupils larger than normal, irregular; react slightly to light. Looks up and down easily, but only with difficulty to either side; can by a great effort perform associated movements. Reaction to light if present at all is very slight. Winks the right eye less often than the left, and when she winks both, the right lid is not closed tightly. Mouth drawn to the right; the right side of the face droops; puffing of the right cheek; tongue protruded to right side; has to be fed; is apparently completely blind; elevates the brow equally on both sides. Right arm and leg are paretic; some spasticity is present, but no contractures; reflexes on this side are prompt. Sensation to pain and touch, apparently normal all over body. Patient very emaciated; Babinski reflex not obtainable. Patient able to move right lower limb better than right upper limb. Each crystalline lens discovered to be cataractous. Chest: Protrusion of sternum. Fine moist rales heard over both lungs. Heart sounds faint and irregular. Slight diastolic murmur in mitral area. Pulse weak, irregular, and of small volume. Arteries greatly sclerosed.

At the autopsy the anatomical diagnosis was: General arteriosclerosis; chronic endocarditis; perisplenitis; chronic interstitial splenitis; chronic interstitial nephritis; cystic kidney; atrophic cirrhosis and passive congestion of the liver; thrombosis in thoracic aorta; hydrops of the bladder.

Microscopic study of the spinal cord and brain showed the following:

Lumbar Region. The cells of the anterior horns show considerable change, of the same character as in the previous case. There is occasionally a peripherally placed nucleus, and here and there breaking up of chromophilic elements. The cellular pigmentation is excessive, at times nearly the whole of the cell being taken up by the pigment. The bloodvessels, especially those in the posterior columns, show thickening. No distinct degeneration of nerve fibers in any of the tracts of the white matter, Not even in the crossed pyramidal tracts.

Thoracic Region. Appearance similar in every way to that in Case I.

Cervical Region. Changes in anterior horn cells same as those in the lumbar region. The thickening of the blood-vessels is not as marked as in the thoracic region. In the posterior columns, especially in their posterior portions, there is an evident sclerosis, the tissue here staining poorly by the Weigert hematoxylin. The sclerosis of the blood-vessels is not as marked as in the thoracic region.

Paracentral Lobules. Most of the cells of Betz are diseased; eccentric nuclei are seen, and many of the cells are diffusely stained and spherical in outline. Pigmentation is not as marked as in Case I. Thickening of the blood-vessels is very evident, both in the pia and within the brain tissue; and beneath the surface amyloid bodies are fairly numerous.

Optic Chiasm and Optic Nerves. Transverse sections of the optic nerves unfortunately were not cut; in the sections through the optic chiasm cutting the optic nerves longitudinally, the latter, besides showing great thickening of their bloodvessels, show many degenerated nerve fibers. Large numbers of amyloid bodies are present in the optic nerves.

Olfactory tract and bulb show amyloid bodies as in Case I.

CASE III.—Bagnall, a woman, aged sixty-two years; occupation, housework. Admitted to the Philadelphia General Hospital, service of Dr. Spiller, March 3, 1906. Patient is weak and somewhat confused. Cannot answer questions intelligently.

History. Two years ago, two strokes of apoplexy. Two weeks ago, caught cold. Very debilitated, weak, and confused. Cannot answer questions intelligently. Old and wrinkled. Gait unsteady. Dr. Weisenburg's notes on her condition: Patient says she is thirty-six; obeys commands. Wrinkles brows equally well on both sides. On shutting eyes, power is apparently equal on both sides. On open-

ing mouth and showing teeth, a weakness of the lower part of the right side of the face is apparent. The tongue is protruded to the right. The power in the patient's hands is diminished, equally so on either side. It is impossible to tell whether or not there is an old paresis on the right side. Biceps and triceps jerks cannot be obtained because the patient holds the arm so rigidly. Patellar and Achilles jerks are prompt on the left side; on the right side they cannot be obtained, as the patient holds her limb so rigidly. Eyes show arcus senilis; pupils normal in size but slightly irregular in outline. React sluggishly if at all to light and accommodation. Extra ocular movements performed fairly well. Tongue moist, broad, protruded in median line. No irregularities in facial lines; chest emphysematous; kyphosis is present in upper thoracic region. Expansion is slight; percussion gives slightly hyperresonant note. Crepitant and subcrepitant rales at the bases. Heart sounds fairly clear. Lower limbs show nothing abnormal; knee-jerks and sensation normal. There is an area of inflammation over the right elbow. Biceps and triceps reflexes normal. Gait very unsteady.

March 30th. Patient's condition is getting much worse. She is very dyspneic, and her chest is full of rales. Heart rapid and weak.

March 31st. Urine: trace of albumin; few coarsely granular casts. Died today.

Autopsy. Anatomical diagnosis; congestion and edema of pia mater; brown atrophy of heart; chronic endocarditis; general arteriosclerosis; esophageal piles; enteroptosis; nutmeg liver; cyanotic induration of spleen; fatty, sclerotic pancreas.

Microscopic study of spinal cord and brain showed the following:

Lumbar Region. There is marked pigmentation in the cells of the anterior horns. The bloodvessels within the cord are thickened. By the Weigert hematoxylin stain the right crossed pyramidal tract is found to be slightly degenerated.

Thoracic Region. A very slight degeneration of the crossed pyramidal tract of the right side is seen. There is a very evident hydromyelia, the dilated central canal extending from the base of one posterior horn across the cord to the base of the other posterior horn. The cavity is lined by ependymal epithelium at certain regions, which seems to be undergoing proliferation and to be extending into the surrounding tissue. The bloodvessels in the gray matter appear congested. Amyloid bodies are present in large number, and the vessels are thickened.

Cervical Region. Most of the cells of the anterior horns are deeply pigmented, and some are entirely filled with pigment. Strands of tissue extending into the cord from the pia, and the walls of the bloodvessels are thicker than normal. There is slight degeneration in the right crossed pyramidal tract. The central canal is a little dilated. By osmic acid staining, some black fibers are seen in each crossed pyramidal tract, more numerous on the left side.

Medulla. More black fibers in right pyramid than in left.

Paracentral Lobules. The cells of Betz in each paracentral lobule are excessively pigmented. The bloodvessels in the pia and in the cortical gray matter are considerably thickened. Amyloid bodies are present, but not in very large number.

Optic Chiasm and Optic Nerves. No degenerations are found in the optic nerves; but each nerve shows a great increase in fibrous tissue. Amyloid bodies are present, especially, as in the other two cases, at the sides of the chiasm.

Olfactory Tracts and Bulbs. Amyloid bodies as in the other two cases.

A small area of softening was discovered in the lenticular nucleus of the one side. Examined microscopically, the tissue here is seen to be very much ossified.

Resume. The pathological findings in the three cases were fairly uniform. The bloodvessels throughout cord and brain were found thickened and at times with lumen obliterated,

this condition being especially marked, in all three cases, in the thoracic segments of the cord.

The connective tissue in the pia was seen to be increased; that between the fibers of the optic nerves was markedly proliferated.

In the nerve cells of the anterior horns and in the cells of Betz in the paracentral lobules there was seen a variety of changes. In Cases I and II, both in the lumbar and in the cervical regions, the cells showed, in addition to intense pigmentation, distinct degenerative changes. Case III showed, in the anterior horn cells, merely an intense degree of pigmentation. The cells of Betz in the first two cases presented the same diseased condition as was seen in the anterior horn cells; in the third case, however, the cells of Betz were, aside from intense pigmentation, unchanged.

Quite remarkable were the enormous numbers of amyloid bodies present in the three cases. These bodies were seen all through the cord, at its periphery, beneath the pia in the cortex of the brain, at the sides of the optic chiasm, and in the olfactory tracts and bulbs.

As for nerve-fiber degenerations, none were present in either of the first two cases. Case III showed, by the Weigert stain, a slight degeneration of the right crossed pyramidal tract throughout the cord; sections stained by osmic acid showed a recent degeneration of the left pyramidal tract.

These cases, though offering nothing distinctly new either in symptomatology or in pathological anatomy, may yet serve as contributions illustrating the various manifestations to which arteriosclerosis may give rise.

The symptoms present in these cases are to be in large part attributed to the pathological changes found in the spinal cord and in the brain. Besides the evidences, both in cord and brain, of the poor nutrition which must have been consequent upon the presence of thickened and sometimes obliterated arterioles, there were, in the first two cases at least, distinct degenerative changes in the cells of the anterior horns and of

the motor cortex. These changes, together with the intense degree of pigmentation seen in the cells, both of which changes were in all probability due to the cutting down of the blood supply, must be held to account for the paraplegia and the contractures in the lower limbs.

Arteriosclerosis has been much studied of late, especially that form of disease confined to the nervous system; and it has come to be recognized that an absence of thickening of the peripheral arteries does not by any means exclude the possibility of the presence, even in a markedly advanced degree, of sclerosis of the arteries of some internal organ. Within the last year a new manifestation of arteriosclerosis of the central nervous system has been described—that known as intermittent claudication of central origin. This disorder, first described by Dejerine,¹ in April, 1906, and later by Grasset,² Sollier,³ and others, differs from the form of intermittent claudication dependent upon peripheral arteriosclerosis—a condition first recognized by Charcot in 1888—in several particulars; the signs of peripheral arteriosclerosis are absent (cyanosis of the part, coldness, loss of pulsation in dorsalis pedis artery); the knee-jerks, which may be normal or even decreased when the patient is at rest, become markedly exaggerated after exercise; the Babinski reflex may be present after exercise; there are generally bladder and rectal disturbances, and perhaps disturbances in sexual function.

In Dejerine's cases the symptoms came on only after exercise; after the patients had walked for varying distances there developed in the lower limbs first a sensation of heaviness, then an actual paralysis. After resting for a while, no muscular weakness whatever could be discovered, and the exaggerated knee-jerks and Babinski's sign which had been present disappeared. Since the peripheral arteries in the lower limbs were comparatively normal, Dejerine attributes the symptoms to an ischemia of the spinal cord, the result of an endarteritis.

Sollier³ believes that the nature of the process is always syphilitic, in spite of the fact that many of those affected denied having had syphilis. An endarteritic process, he says, brings about an insufficient blood supply to the dorso-lumbar region of the cord, and so produces the symptoms.

Erb also emphasizes the distinction between the central and peripheral forms of arteriosclerotic lameness, and reports a typical case of the central type with relatively normal peripheral arteries.

Besides this form, various other symptom complexes, the result of arteriosclerosis of the nervous system, are reported in the literature. Thus, Collins gives an instance in which the symptoms came on so rapidly that the case was looked upon as one of Landry's paralysis. Buzzard and Barnes⁴ report a case of a woman of fifty-three years, who had suffered for ten years with a gradually progressing weakness in the legs and the facial muscles; also from uncontrollable laughter. At autopsy, degeneration of the pyramidal tracts was found, which the authors attributed to the manifest disease of the arterioles. The cases reported by Pic and Bonnamour⁵ showed a course characterized by "paretospasmodic" symptoms, exaggeration of reflexes, presence of the Babinski reflex, epileptoid trepidation, and weakening of intellect.

Hirsch⁶ found, in a number of cases of "arteriosclerosis of the spinal cord," besides thickening and partial obliteration of the lumina of the arterioles, pathological changes in the anterior horn cells, and also minute hemorrhages into the gray matter of the cord. Such hemorrhages are also described by Warrington and Owen,⁷ and are believed by them to be due to obliterative arterial disease. On the other hand, in the case reported by Wilson and Crouzon,⁸ and called by them "senile combined sclerosis," in which there was very evident sclerosis in the posterior and lateral tracts, there were no marked vascular changes, and no anterior horn cell lesions.

Burr,⁹ in 1904, emphasized the fact that many of the cases

of partial palsies, vague rheumatic pains, and other disturbances of senility, have their probable origin in central lesions due to the presence of diseased bloodvessels.

Naka, however, in a careful study of the pathological findings in 17 cases of senile dementia, states that he does not consider that the symptoms of spasticity, tremor, and increased reflexes have a sufficient explanation in the few changes which he was able to detect in the spinal cord and brain. He describes, however, thickening of bloodvessels, perivascular glia proliferation, and pigmentation and vacuolation of nerve cells.

That we are dealing, therefore, in cases such as those here reported, with symptoms or lesions the result simply of a deficient blood supply to the cells and fibers of the brain and cord, is most probable. Why the arteriosclerotic process should pick out in certain individuals the peripheral arteries, and in others only the central arteries, is as yet unexplained.

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POST-APOPLECTIC TREMOR (SYMMETRICAL AREAS OF
SOFTENING IN BOTH LENTICULAR NUCLEI AND
EXTERNAL CAPSULES.)*

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The features of interest in the case to be reported are, clinically, a tremor of the right arm, resembling post-hemiplegic tremor, and ataxia of the arms and legs, and, pathologically, the presence of symmetrical lesions of the putamen.

The patient, a man of 58, was admitted to the Philadelphia Hospital on April 20, 1903, his chief complaint being difficulty in walking. The family history, as well as the previous history, were unimportant. Owing to the mental condition of the patient, a reliable account of the onset of his disease was not obtainable, his answers being frequently contradictory.

He stated when admitted that his present trouble had begun 3 years previously. He complained of pain in the head, weakness and numbness of the legs, and weakness of the right arm. At this time nothing of moment was noted excepting that the right knee jerk was increased, some ataxia was present in the upper limbs, and the patient had a tendency to lose his balance if he turned suddenly while walking. There was no Babinski reflex, and no apparent weakness in any of the limbs.

He came under the care of one of us (Dr. Potts) in April, 1906. At that time the peculiar movements of the right arm attracted attention. These consisted of alternate flexion and extension at the elbow and wrist, the hand being supinated. The movements were almost constant, only ceasing for a time if his attention was distracted. At this examination he told a somewhat different story than at first, saying that at the beginning of his disease he noticed weakness of the legs, the left being weaker

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than the right, and that these symptoms dated back eleven years, following what he called "a cold." Six years later the right arm became affected, the disorder consisting at first of a difficulty in writing. Examination at this time showed a somewhat ataxic gait, which was more apparent with the shoes off. The right leg was held more stiffly than the left. Station, with the eyes closed, was poor, and there was also marked ataxia of the arms, especially the right. There was apparently no weakness of the arms. The



Fig. 1. Linear area of softening in external capsule on right side. Upper limit. (Macroscopical section.)

legs appeared to be somewhat weaker than normal. The biceps, triceps, and wrist jerks were not marked. The knee-jerks were prompt, the left being more so. The plantar reflex gave a normal response. Examination of the eyes showed a sluggish response to light, but otherwise they were normal.

The brain and spinal cord were turned over to Dr. William G. Spiller, who gave them to one of us (Dr. Rhein) for examination. The specimens were hardened in formalin.

Horizontal sections of the brain on the right side revealed the presence of an area of softening which first made its appearance a short distance above the lenticular nucleus, and was situated in the external capsule, *i. e.*, between the claustrum and the internal capsule. This area of softening was linear in shape, and measured at this level 2.5 cm. in length, and about 1 to 2 mm. in width. While it lay directly external to the fibers of the internal capsule, it did not cause any degeneration of these fibers.



Fig. 2. Area of softening extending into the middle portion of the right putamen (microscopical section, section reversed in photograph).

The focus extended downward in a vertical direction to a position corresponding to the lower limit of the island of Reil. At the lower level of the lenticular nucleus it destroyed in part the external portion of the putamen in its posterior third. At the level at which the fasciculus of Türck is seen ("temporale Brückenbahn"—Obersteiner) these fibers were in part destroyed. The anterior commissure was intact, as were also the fibers of the

internal capsule. The fibers of the external part of the foot of the peduncle, corresponding to the situation of the fasciculus of Türk, stained poorly. The red nucleus appeared to be intact.

On the left side of the brain there was a similar lesion which began at a somewhat lower level than the one on the right side, its upper limit being at about that level where the lenticular nucleus is first seen in horizontal sections. It was situated in the same relative position as that of the cavity on the right side, occupying the region of the external capsule between the claustrum and the putamen, and measured 3.5 cm. in length, and 10 mm. in its widest portion. It extended downward in a vertical

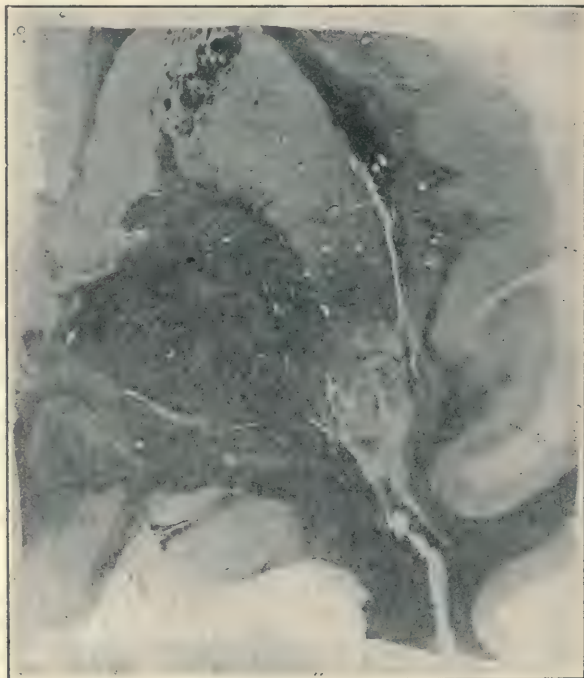


Fig. 3. Area of softening in the posterior portion of the left putamen (microscopical section, section reversed in photograph).

direction, and disappeared a little above the level of the lower border of the island of Reil. The posterior third of the putamen in its lower part was in large measure occupied by this focus of softening. The fibers in the retro-lenticular region were more or less involved.

The fibers in the internal capsule were not degenerated. The red nucleus appeared to be normal.

Certain fibers passing inwardly and apparently through the lenticular nucleus toward the thalamus, stained well on both sides.

In sections from the pons, medulla oblongata and spinal cord the nerve fibers stained normally. There was no evidence of descending degeneration in the pyramidal tracts.

The cells of the anterior horns in the cervical and lumbar regions, stained by hemalum and acid fuchsin, showed no marked change.

Sections from the cerebellum showed an unusual vascularity of the dentate nucleus on both sides. The vessels were numerous and somewhat thickened, and surrounded by a slight round-cell infiltration, and there was also some fresh hemorrhage about some of the vessels.

To summarize the pathological findings in this case, there was an area of softening in each putamen, which caused degeneration in the fasciculus of Türck, on the right side. There was no degeneration in the motor fibers of the internal capsule, the pons, medulla oblongata or spinal cord.

Associated with these pathological findings, the tremor of the right arm and the ataxia of the arms and legs are worthy of some discussion.

In regard to the first of these symptoms, namely, the involuntary movements of the right arm, which resembled a post-hemiplegic tremor, it is interesting to note that lesions of the lenticular nucleus have been thought by some to cause post-hemiplegic chorea. Among the first to describe cases of this nature was Demange,¹ who, in 1883, reported 11 cases with autopsy, in 6 of which the lenticular nucleus was the seat of disease.

Sander,² in 14 cases of athetosis collected from the literature, found the corpus striatum involved in 7 cases. In the case described by Grasser and Rauzier³ the lenticular and caudate nuclei, and the internal capsule were partly destroyed by an area of softening.

The putamen was symmetrically diseased in one case described by Hebold,⁴ and in a second case the claustrum was also involved on both sides. He quotes Nothnagel who collected cases of athetosis and hemichorea, in which the lenticular nucleus and the internal capsule were involved. In Anton's⁵ case the choreiform and athetoid movements were on the same side as the lesion in the putamen, and in Jacob's⁶ case the contralateral lenticular nucleus was the seat of the disease.

Many cases have been cited in which, besides the lenticular nucleus, the thalamus has also been involved. This was true in 50 out of 78 cases collected by Bidon.⁷ In about 100 cases studied by v. Monakow,⁸ in 70 per cent. to 80 per cent. there was also lesion of the internal capsule.

On the other hand, bilateral lesions of the lenticular nucleus may occur without causing tremor, as in Edinger's⁹ case, and in 2 cases of Hébold.

If foci of softening or other lesions of the lenticular nucleus cause tremor, it is hard to reconcile entirely the findings in our case with this statement, as tremor was only present in the right arm, while the putamen was diseased on both sides. Of interest in this connection is the opinion of Hébold who believed that a different explanation for the motor disturbances than injury to the putamen, must be looked for, at least as far as the outer portions are concerned.

It should be remembered, however, that in our case the lesion caused much greater destruction in the left putamen than in the right, involving, in the former, a large part of the posterior third of the putamen, while on the right side the lesion was linear in shape and occupied the external portion, the destruction of the putamen being less extensive. It may be that the lesion on the right side was not sufficiently large to cause disturbances of motion, while the damage to the left putamen, being much more extensive, was sufficient to give rise to a tremor in the right arm. This is of course somewhat theoretical.

At least the findings in our case permit the statement that bilateral lesions of the putamen do not always cause bilateral disturbances of motion.

The pathology of post-hemiplegic disturbance of motion has not been definitely established, and the views held upon the question are far from uniform. On the whole, however, the weight of evidence is in favor of placing the lesion in one of several localities, and preferably in the thalamus, or in the pons, in the neighborhood of the superior cerebellar peduncles and red nucleus. Many observers have, however, described tremors, choreiform movements and athetosis post-hemiplegic in type, in organic disease of the cerebellum, medulla oblongata, cortex and spinal cord.

Lesions of the thalamus have been regarded by many observers as the chief cause of these movements. Since Charcot,¹⁰

in 1873, reported 3 cases with autopsies, in which the lesion involved the posterior portion of the optic thalamus, caudate nucleus, and corona radiata, a number of cases of this character have been described, namely, those of Stephan,¹¹ Sander, Gowers,¹² Bidon, Kahler and Pick,¹³ Raymond,¹⁴ Bischoff,¹⁵ and Leyden,¹⁶ who found various lesions affecting the thalamus, with or without involvement of the surrounding tissues.

In 100 cases studied by v. Monakow, it is stated that, in the majority of cases the lesion was found principally in the thalamus. In another group of cases the retrolenticular portion of the internal capsule and the thalamus were the seat of disease, while in the remainder of the cases the posterior portion of the corona radiata and the lenticular nucleus were involved.

Raymond, however, on the contrary, noted in 35 cases of hemiplegia, involvement of the thalamus in only 4 cases, and Sander believed that there were only a few cases in which the thalamus alone was involved. Moreover, the thalamus and the internal capsule were affected together in 20 cases in the literature studied by Stephan.

The relation of the post-hemiplegic movements to the cerebellar peduncles, the tegmentum, and the red nucleus, was probably first described by Benedickt¹⁷ who, in 1874, reported 22 cases of hemiparesis and chorea caused by a lesion in the crus cerebri, at the height of the oculomotor nucleus. Halban and Infeld¹⁸ added to these 2 cases of their own, in one of which a tuberculous focus had destroyed the red nucleus in large part, extending almost to the decussation of the superior cerebellar peduncles. Henoch¹⁹ also observed a case with tremor, hemiplegia, and crossed oculomotor palsy, in which there was found a tuberculous lesion of the pons and corpora quadrigemina, and Bergen²⁰ collected 10 cases in which the cerebellum and superior cerebellar peduncles were destroyed, causing incoördination identical with hemichorea on the same side as the lesion.

The superior cerebellar peduncles, the red nucleus and the tegmentum were diseased in a case reported by Bonhoffer,²¹ who believed that choreiform movements and athetosis were due to a lesion of the centripetal fibers extending from the cerebellum to the cerebral cortex, giving rise to a functional disturbance of the motor cells of the cortex. He advanced three reasons for accepting this view, namely, (1) the constant presence of a lesion of

the superior cerebellar peduncles, or its projections into the sub-cortical ganglia; (2) the hypotonia of the musculature; (3) the disturbance of voluntary motion in chorea. He thought it probable that, in consequence of a more or less complete interruption of the fibers, the centripetal impulses only in part reached the cortex, while others passed along the centrifugal fibers and gave rise to automatic movements the character of which depended upon the nature and extent of the lesion.

Pineles²² reported 2 cases of athetosis which he considered of cerebellar origin, and cited the case of Oliver, in which there was a glioma of the vermiform process in a case presenting tremors of the arms and legs. He cited also Menzel's case of choreiform movements with atrophy of the cerebellum, a similar case of Meynert's, Ceni's case of atrophy of the right hemisphere of the cerebellum, and right cerebellar peduncle, with a hemorrhagic focus in the left red nucleus, and Sander's⁴ case in which there were choreiform movements on the right side, caused by a gliosarcoma in the right cerebellum, destroying the corpus dentatum. Pineles concluded that in many cases choreiform movements and athetosis were caused by disease of the cerebellum, or the cerebellar peduncles. Other cases of a similar nature have been reported by Kirschhoff,²³ Bonhoffer, Huppert,²⁴ Muratoff,²⁵ and Hammarberg.²⁶ In Hammarberg's case the movements were pendulum-like.

Lesions in the medulla oblongata, causing hemichorea, were described in Bidon's cases (collected from the literature), in v. Monakow's case and in the cases of Fropier,²⁷ and Broadbent (in v. Monakow) and Henoch.

Cortical lesions have been described as causing athetoid and allied movements in the 2 cases of Demange, and those of Balfour,²⁸ Beach,²⁹ Korella,³⁰ and Hudovernig.³¹ Some of these cases are, however, so meagerly reported as to be of little value.

Opposed to this is the statement of Exner (in Frey) who collected 167 cases of cortical lesion without one instance of post-hemiplegic tremor. In this connection, as bearing upon the irritation of the motor tracts as causing post-hemiplegic movements, must be mentioned the cases in which the internal capsule has been the seat of the lesion, as illustrated by the case of Demange, and those collected by v. Monakow, Bidon (29 cases), and others.

Finally, Eisenlohr³² and Anton describe cases in which spinal

lesions were mentioned as causing the movements under discussion.

Several theories have been advanced to explain the occurrence of these movements, among the first of which was that of Charcot, who believed that they were due to disturbances of a "chorea-bundle," a view long since abandoned.

Kahler and Pick, who concluded that cysts or areas of softening in the thalamus, with or without involvement of the internal capsule, give rise to hemichorea and other disturbances of motion, believed that these were due to irritation of the pyramidal fibers, a view also held by Sander,²⁹ Demange, Kolisch,³³ Hudovernig, and Nothnagel.³⁴ Frey⁴⁰ and others, however, object to this theory on the ground that the usual result of irritation of the pyramidal tracts is to cause convulsions and spasms; that hemiplegia is a frequent occurrence, while choreiform movements are rare; that lesions removed from the motor tracts cause these symptoms; that paresis is absent in some cases; that it is improbable that complicated movements should remain circumscribed if thus explained (v. Monakow); that the head is affected in deep-seated lesions, and that these symptoms are infrequent in lesions of the lenticular nucleus (Frey). Furthermore, it is advanced by some authorities that cortical lesions rarely cause post-hemiplegic movements, although the cases of Demange controvert this statement.

Von Monakow does not subscribe to the view held by Kahler and Pick and others, that these movements are the result of irritation of the pyramidal fibers which is conveyed to the cells of the anterior horns of the cord, but in his opinion the tissues surrounding the lesion, as in the subthalamic area, the tegmentum and the pons, become irritated, and that this irritation is transmitted to the motor cortex.

Bonhoffer, in explaining the symptoms, believed that the different choreiform movements are caused by alterations of the impulses which normally pass to the cerebral cortex through the tegmentum. As already stated, he believed that it was plausible that impulses pass centripetally to the cerebral cortex in part only while others pass directly in the centrifugal motor fibers, and give rise to automatic movements.

According to Frey and Stephan, the thalamus is a coördinating center which, when disturbed, gives rise to post-hemiplegic disturbances of motion. Frey believed that the cause of these

movements is to be found in lesions of the thalamus and hypothalamic region.

Anton held that automatic associated movements originated in the posterior part of the optic thalamus and its connections, while the corpus striatum inhibited these movements. Therefore, lesions of these ganglia cause a lowered inhibition, and in consequence an increase of the automatic movements, or chorea.

Halban and Infeld believed that fibers going through the red nucleus have to do with a complicated mechanism, and that disturbance of these fibers gives rise to the automatic movements.

According to Sander, disease of the thalamus was the cause of athetosis in his case, and he expressed the opinion that lesion of the motor tracts causes in part of the fibers incomplete interference with the passing of nerve impulses, and that the accumulation of these impulses centrally from the lesion, *i. e.*, in the cortical cells, becomes so intense from time to time as to overcome the resistance at the point of the lesion, giving rise to movements of a rythmical nature.

Pineles inclined to the belief that while these movements may be associated with irritation of the pyramidal fibers, many of the facts speak for the relation of chorea to the superior cerebellar peduncles. He believed that the choreiform movements were the result of loss of function (an "ausfall" symptom).

One is struck, in studying the cases and the various theories above cited, with the fact that the majority of lesions described affected centers or fibers which are connected directly or indirectly with the cerebellum, and it does not seem improbable that, in some way not perfectly clear, the function of the cerebellum, at least as far as co-ordination is concerned, may be disturbed, and, as a result, give rise to choreiform movements, athetosis and tremor, which, according to some observers at least, may be looked upon as allied disturbances of motion.

The findings in our case may add weight to this view, if we are correct in assuming that the lenticular nucleus is associated with the cerebellum by fibers which pass from the lenticular nucleus on one side, to the contralateral cerebellar hemisphere by way of the ansa lenticularis.

Obersteiner³⁵ believes that the lenticular nucleus is connected with the lower olive on the same side, and the opposite cerebellar hemisphere in this way, and Mills³⁶ states that the corpus striatum

is connected with the cerebellum through the red nucleus, the pontine nuclei, and olives, and the other basal ganglionic deposits.

According to Dejerine³⁷ part of the fibers of the ansa lenticularis penetrate into the antero-internal part of the capsule of the red nucleus, which is composed of fibers from the superior cerebellar peduncles.

These facts justify the assumption, to some extent, that the lenticular nucleus may have some function relating to co-ordination through its connections with the cerebellum which it may have at least indirectly, if not directly, by reason of its relation with the red nucleus. If, then, as a result of a lesion of the lenticular nucleus, the fibers associated directly or indirectly (through the red nucleus or otherwise) with the cerebellum be interrupted, and thus the functions of the cerebellum be impaired, may not disturbances of motion arise expressing themselves in involuntary movements or ataxia?

This brings us to the consideration of the ataxia of the arms and legs which was present in our case, and offers a possible explanation for this symptom, which is not explained in any other way by the pathological findings already described.

The tremor in our case, we believe, was not hysterical, as the patient exhibited no other stigmata of hysteria, although of course tremor may be the only manifestation of hysteria, as in the case described by Mitchell and Spiller.³⁸

IN CONCLUSION, we believe:

1. That the lesion in the left lenticular nucleus may have been responsible for the tremor of the right arm in our case, and the association of unilateral tremor with bilateral lesion may be explained by the fact that the lesion in the right putamen was not extensive enough to set up sufficient irritation to cause this symptom on the left side.

2. That post-apoplectic disturbances of motion may be due to lesions of the lenticular nucleus, the optic thalamus, and the pons in the region of the superior cerebellar peduncles, and of the cerebellum, the cause being, in all instances, a disturbance of co-ordination. When the lesion is extra-cerebellar the cause of this disturbance is possibly an indirect result of destruction of fibers related to the cerebellum directly or indirectly through the red nucleus.

3. That the ataxia in our case may have been indirectly of cerebellar origin.

It must not be forgotten, in drawing conclusions from an analysis of the above cited cases, that there are on record instances in which lesions of the posterior portion of the thalamus ("hintere Sehhügelgegend"), red nucleus, superior cerebellar peduncles, dentate nucleus, and cerebellum have occurred without causing athetoid and choreiform movements (v. Monakow).

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PERIPHERAL SPINAL DEGENERATION RE-
VEALED ONLY BY LONGITUDINAL SEC-
TIONS OF THE CORD AND AN
AXIS CYLINDER STAIN.

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This woman, aged forty, and married, had an illness of three weeks which began with inflammation of one joint, followed by inflammation and tenderness of another, the pain leaving one joint as it attacked the next. Her temperature ranged about 103° F., pulse 120, and as she grew steadily worse the pain became very intense, with joints swollen and red.

The patient had no disturbance of motor function or change in sensibility indicating involvement of her nervous system. Three days before her death she became delirious and had to be restrained; she said during the course of the delirium that she had been drinking prior to admission, but this dated two days before and could hardly be the cause of the delirium. The fact of her history of drinking suggests the possibility of a gonorrhoeal infection, though that does not necessarily follow. She developed complete suppression of urine after the delirium and remained in this condition until her end.

The notes taken at the autopsy are: "Pachymeningitis

Ludlum: Peripheral Spinal Degeneration.

and leptomeningitis, and œdema of the brain; acute nephritis in addition to chronic nephritis, fatty infiltration of liver, acute splenitis, subpericardial hæmorrhages, fatty degeneration of heart, subpleural hæmorrhages, with congestion and slight œdema of lungs; chronic aortitis. Dura adherent throughout the cranium. -Pia, whitish, especially over occipital and parietal lobes."

A microscopical examination of brain and cord showed a moderate degree of meningitis, especially of the cord. There were no cultures taken from meninges at the autopsy, so we are forced to judge from the clinical symptoms, which would seem to indicate a rheumatic or gonorrhœal affection of the joints, the same ætiological factor extending into the spinal membranes. Microscopical sections in hæmatoxylin and eosin showed a meningitis about the entire periphery of the cord extending somewhat into the longitudinal fissures, but limited in amount in these parts. There was a moderately thick, peripheral, fibrinous accumulation, within which there were many round cells.

The interest of the case centres about the fact that there were no lesions within the cord demonstrable with the Weigert-Pal stain by cross section, but with the Bielschowsky stain for neurofibrils when the sections were cut longitudinally there was a band of degeneration and breaking up of axones about the entire periphery of the cord. Marchi stains showed nothing.

It is fair to assume from the normal condition of the nerve roots on examination that the cord condition was due to the meningitis; there was no infiltration of leucocytes in the degenerated area and the condition would seem to be a toxic change.

The nerve cells in the cord were markedly altered in all groups of cells in the anterior horn. There was no congestion of arterioles in the gray matter, and the changes noted in the cells were those of an acute change, that is, a fine granular breaking up of neurofibrils. There was, in

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addition, a deposit of yellowish pigment in the majority of the cells, due to the life of the patient having been a hard one and hence showing the results in a presenile condition of the cells in the cord.

The cells in the cortex, notably the Betz cells, were in some instances normal, and others were markedly degenerated. All showed a more or less accumulation of yellow pigment, and those degenerated showed an acute breaking up of neurofibrils. These changes were sufficient to cause the mental change observed in the delirium, though it must also be taken into consideration that the changes might have been produced by the fever.

The effort to examine the intermediolateral column and the column of Clarke was without decisive results, as all the cells in the gray matter in the cord were changed in equal amount.

We have in this case a process of degeneration encircling the cord varying in depth and probably occurring as a result of contiguous meningitis. The variation in depth showed the greater amount of degeneration in the region of Gowers's tract, and this is the region shown by Marie to be the place of anastomosis of the posterior and anterior vessels of the cord.

Marie advocated the opinion that system diseases of the cord were delineated by the anatomy of the circulation and were not necessarily neurone system diseases. The fact that, with a stain coloring axis cylinders, the greatest degeneration to an influence equally surrounding the cord was in the region of anastomosis, and that this is the region noted by Marie as the boundary of the degeneration

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found in posterior lateral sclerosis is worthy of notice, for it shows that either the anterior or posterior arteries being the seat of disease, the anastomosis would be barely sufficient to overbalance any deficiency in nutrition in the diseased portion and in toxic states of the cord that is the part most affected.

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